DIAGNOSIS IN LOCOMOTOR DISORDERS

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PREFACE

This book is a series of articles on differential diagnosis in a number of prominent symptoms and signs in disorders of the locomotor system. These are the diagnostic problems presented to those who work in physical medicine departments and rheumatic units of general hospitals and they are part of the daily work of all general practitioners.

They are often difficult problems their solution may be found in so many different fields of medicine. I know of no other book on the diagnosis of diseases that present as a disorder of the locomotor system

My aim has been to provide practitioners with a short book restricted to essentials, for quick reference. The presenting symptoms or signs whose diagnosis is discussed are arranged alphabetically If what the reader wants cannot be found promptly from this arrangement, a reference to the index should locate it.

In a book of this structure some repetition is inevitable I have tried to reduce this to a minimum by giving the main account of a disease under its most common presenting symptom Whenever the disease has to be mentioned elsewhere, a reference to this main account will be found.

K.S

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ACHING, GENERALIZED

I shall exclude those diseases in which general aching is but an incident in a more important symptom complex, such as acute specific infections severe rheumatoid arthritis, and the like. The complaint is often aching all over Inquiry usually clarifies this to never free from aching somewhere —that is, a transient and vagrant aching. Less often it is continual and widespread Aching may be felt in joints, or more diffusely

Data needed for diagnosis are provided by the history and by a routine clinical examination, which includes the spine bones and joints. Unless the cause is recognizable without difficulty—as it would be in paralysis agitans [p. 61] or in acromegaly [p. 30]—it will usually be wise to get X ray films of the spine, a blood count, and an erythrocyte sedimentation rate (E.S R.) estimation. Other X ray and laboratory studies may be indicated

The differential diagnosis is

JOINTS

Spondylosis

Polyarticular osteoarthritis

Arthralgia without physical signs in young people

pre rheumatoid state pre-spondylitic phase of ankylosing spondylitis, subacute rheumatism.

BONES

Metabolic rarefying diseases of bone

Paget's disease

Acromegaly

SOFT TISSUES

General debility and malnutrition

Painful subcutaneous fat

Fibrositis

Paralysis agitans

JOINTS AND BONES

Referred pains from the multiple lesions of spondylosis [p 97], or of polyarticular osteo-arthritis [p 189] are easily recognizable Vagrant arthralgia in young people is a more difficult problem, and needs especial care [p. 195]

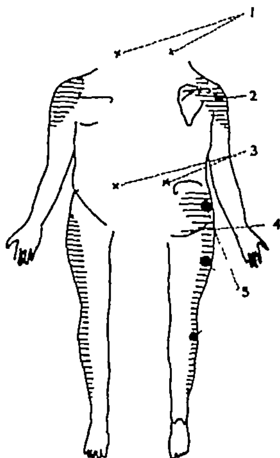


FIG. 1

Distribution of referred pain from experimental pain stimuli applied to interspinous ligaments C.4-5 and L.5-S.1 1 interspinous lig. C.4-5 2. area of referred pain C.5 3 interspinous lig. L.5-S.1 4 area of referred pain L.5 5. referred tender spots (after Kellgren).

The presenting symptom in the metabolic diseases of bone [p 31] is likely to be pain in the back and limbs, often ascribed by the patient to 'rheumatism' The need for X ray examination of the skeleton may be suggested by a spontaneous fracture, or by the appearance of bone deformity The first complaint in Paget's disease [p. 28] may be of a diffuse deep aching in back and lower limbs. The disease will be discovered by routine radiography of the lumbar spine and pelvic bones.

SOFT TISSUES

General debility and malnutrition

In general muscular weakness (of whatever origin) ordinary activities will evoke widespread muscular aching, just as transient aching follows

prolonged exertion in normal subjects. In the serious cachexias it will be a comparatively minor complaint It is prominent when debility is due to less serious causes chronic infections, such as cystitis or nasal sinusitis chronic pelvic disorders in women inadequate diet fatiguing occupations and unhealthy living conditions. Any form of anaemia is an important cause of general aching—an aching possibly related to muscular activity under conditions of deficient oxygenation

Painful subcutaneous fat

Panniculitis names a condition fairly common in middle aged women whose weight is increasing. Subcutaneous fat especially on the lateral side of the thighs, around the knees, and on the back of the neck and shoulders, is tender the overlying skin feels thickened and may have an orange peel appearance. The complaint is of a variable aching.

Localized and symmetrical painful fat masses, mainly in the limbs, are a feature of adiposis dolorosa, a name denoting probably several forms of endocrine obesity Tender fat deposits also occur in those forms of endocrine obesity of the lower limbs known as lipomatosis, or lipodystrophy and in some obese cases of Graves disease.

Fibrositis

The commonest diagnostic label for unexplained aching, local or general, is fibrositis. Few now think that it names a single disease process. It is used to denote a number of obscure disorders and I shall try here to see if they can be more clearly defined.

Essentially fibrositis is pain of a peculiar kind a diffuse pain with ill-defined boundaries, and within the painful area several points tender to finger tip pressure. Now these are the characters of referred somatic pain. Experimentally pain excited in a small zone in deep lying muscle, or ligament, or periosteum, is felt diffusely over a large area. When severe, it is referred to a segmental area—a region innervated by the same spinal segment as the tissue stimulated In the painful area are scattered points of deep tenderness [FIG 1] Clinical examples of this experimental phenomenon are common We must therefore begin an investigation of what we feel tempted to call fibrositis by looking for possible sources of referred pain.

There are some who assert that all fibrositis is referred pain, and that the causative lesion is nearly always in the spinal column There may however be additional peculiarities, either in the pain or in associated symptoms, which suggest that the disorder may be a diffuse disturbance of the soft tissues, as we used to think. If no disorder of the spine can be demonstrated there are four and only four possibilities

1 pain is referred from visceral disease

2. there is a spinal lesion though we cannot demonstrate it

3 pain is referred from a soft tissue lesion, such as a sprained ligament

4 referred pain is not the explanation

There are two clinical pictures that seem to merit further inquiry in that they present features that are not part of referred somatic pain

Virus Myalgia. Severe lumbar pain is a well-known symptom of a number of acute virus infections, such as influenza, variola, poliomyelitis

Bornholm disease or epidemic myalgia, has been proved to be an infection by one of the Coxsackie viruses. It is the only proven instance of myalgia due to infection with a myotropic virus, but it seems highly probable that similar viruses are responsible for minor epidemics of acute fibrositis of short duration. In one that has been recorded 41 per cent. of the patients gave a history of a common cold (a virus infection) at the onset of the myalgia.

Myalgia of Autonomic Disturbance The second condition that seems to me distinguishable from referred pain, I think we might still call muscular rheumatism Like referred pain it is a diffuse ache, with localized zones of deep tenderness in the painful area. But this is not all. There is a striking variability pain waxing and waning in severity and extent. There may be relative freedom for several days at a time and indeed some patients are pain-free for many months in the warm weather Pain and stiffness of the muscles are aggravated by rest, and partially relieved by moderate exercise. Pain is sharply aggravated by certain climatic conditions notably a cold damp atmosphere the approach of thunder-storms a sudden change in the weather Draughts are hated, because they bring a highly unpleasant cold numbness to the painful area—e.g. shoulders or sacrum. Attacks may be precipitated by local chilling, by physical fatigue, and by emotional disturbances.

This clinical picture is very well known but there is no kind of agreement as to its explanation. I will merely mention here that it is very commonly associated with one or more of the somatic symptoms of anxiety neurosis complaint of now one, now another of the many symptoms caused by disturbance of the autonomic nervous system. They are due to transient excessive parasympathetic or sympathetic activity and include abdominal symptoms from irregular spasm of smooth muscle, such as the low abdominal aching and discomfort with constipation or diarrhoea from spastic colon tachycardia discomfort in the region of the heart flushes night sweats easy chilling, and cold

blue hands and feet. Autonomic disturbance, by its effect on vascular control, is a sufficient cause of muscular aching and stiffness

Such autonomic symptoms are very often entirely due to anxiety neurosis. But I do not think that this should be accepted as the diagnosis unless there are also the mental symptoms of this complaint a mental state of prolonged fear and anxiety—the patient's thoughts wholly occupied by the sinister portent of his symptoms and other gloomy forebodings—to a degree that has robbed him of his power of con centration, and made him irritable and sleepless. For autonomic disturbance may be entirely of organic origin It is a common result of some chronic toxic or infective conditions, and especially of certain endocrine disorders It is, for instance, seen clearly in both hyper and hypothyroidism, and very commonly in the disturbance of the menopause.

Psychogenic Pain. The muscular aching and stiffness of anxiety neurosis are not imaginary But there is a very different kind of psychogenic pain. There is never the slightest doubt about the diagnosis from the moment these patients begin to speak. It is not a matter of exaggeration—few of us never use superlatives—but rather that nothing of what they say rings true.

They say they have endured very severe pain for months, perhaps years and we are startled to find that this information evokes in us not the least sympathy Pain has been constant, day and night, and nothing has done any good and they say this with a look that conveys a lurking satisfaction rather than suffering. Unlike the genuine pain associated with an anxiety state, their pain never varies. It is continually and uniformly severe but in spite of this awful suffering the patient looks well enough, and can take part in a number of enjoyable pursuits. Their response to examination is equally false. Normal palpation evokes resentful cries of protest, and a writhing withdrawal. It seems to me probable that, whereas the anxiety neurotic suffers genuine aching and discomfort, differing in no way from muscular rheumatism from other causes, the hysterical patient is a subconscious malingerer and has no pain at all.

ATAXY

Disturbance of co-ordination of the several muscle groups taking part in a simple voluntary action results from 1 loss of postural sensibility, 2. cerebellar lesions.

THE CAUSES OF ATAXY

A minor degree of inco-ordination may be seen in many instances of brachial neuralgia from nerve root compression and it results in a clumsiness of the fingers and a tendency to drop things. Temporary ataxy is seen in drug or alcoholic intoxication. There is a hysterical ataxy in which no signs of organic nervous disease will be found.

The causes of ataxy due to organic nervous disease are

SENSORY ATAXY

Tabes dorsalis
Multiple peripheral neuritis
Subacute combined degeneration
Some cases of capsular hemiplegia

CEREBELLAR ATAXY

Thrombosis of a cerebellar artery
Tumour of the cerebellum
Progressive cerebellar degeneration

COMBINED SENSORY AND CEREBELLAR ATAXY

Disseminated sclerosis
Friedreich s disease

SENSORY ATAXY

With loss of afferent impulses from muscles, tendons and joints, which give sense of position and of degree and direction of movement, there results an ataxy that can to some extent be made good by vision

The patient cannot identify objects held in the hand from their shape and size (astereognosis) Told to place a finger tip on the nose with eyes closed, he misses the mark by several inches. Told to stand with arms outstretched, then to close his eyes, the arms gradually fall down. Romberg s sign is positive. With eyes closed the patient cannot confidently describe the position of a toe which you have moved into flexion or extension. The gait is ataxic he walks carefully watching the ground, or may lift his feet too high, to get the feeling of having lifted them at all

In tabes dorsalis [p. 165] an ataxic gait is a late symptom at first it is noticeable only in the twilight.

A purely sensory ataxy is found in subacute combined degeneration [p 188] it soon appears, as an unsteady gait and clumsiness of the fingers The flaccid type may at the onset be indistinguishable from multiple peripheral neuritis [p 14]

CEREBELLAR ATAXY

Closing the eyes does not increase the ataxy and the signs of loss of postural sense are absent. There is nystagmus a staccato ('scanning') speech intention tremor and the cerebellar broken movement in writing or drawing and the cerebellar gait—the swaying staggering gait of a drunken man

Pure cerebellar ataxy is uncommon In contrast to the sudden onset of symptoms in thrombosis of a cerebellar artery and to the rapidly increasing signs of intracranial pressure in the cerebellar tumours that are not uncommon in childhood is the clinical picture of a group of cases of progressive cerebellar degeneration of unknown origin All are very rare Some of them are hereditary symptoms beginning in early middle life Others with no evidence of hereditary origin begin in late middle life, with gradually increasing defect of speech cerebellar ataxy and muscular hypotonia. Subacute cerebellar degeneration has been reported associated with carcinoma especially of the bronchus and of the ovary

COMBINED SENSORY AND CEREBELLAR ATAXY

Associated with sensory ataxy in varying proportions, cerebellar ataxy is seen in disseminated sclerosis [p 212]

Friedreich s ataxia is the only member of a group of disorders known as the hereditary ataxias which is not very rare. Symptoms usually begin between the ages of 5 to 15 years boys and girls being affected equally They are slowly progressive and life is shortened The essential change is a neuronic degeneration of posterior columns pyramidal tracts, and spino-cerebellar tracts. The first symptom is an ataxic gait, the ataxy being both sensory and cerebellar Later ataxy affects the upper limbs, nystagmus is usual and speech is affected. Because both pyramidal and posterior columns suffer weakness of the lower limbs is associated with either slightly spastic or hypotonic muscles, as one tract is more affected than the other The plantar response is extensor and the tendon jerks absent.

Pes cavus and scoliosis nearly always accompany the nervous disorder

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ATROPHY, MUSCULAR

A general muscular atrophy occurs as part of a general wasting it may be due to one of a large number of wasting diseases. The more important to bear in mind are carcinoma, tuberculosis diabetes, hyper thyroidism and anorexia nervosa

The diagnostic problem however is usually concerned with *atrophy of a localized group of muscles*

First find out if the wasting has been accompanied by pain or paraesthesia, and test for sensory changes. The many causes are best grouped in this way

WITHOUT SENSORY CHANGES
Arthritic muscular atrophy
Disuse atrophy
Due to a lesion in the anterior horn cells
poliomyelitis motor neurone disease
Due to primary disease of the muscle
the muscular dystrophies dystrophia myotonica.
Peroneal muscular atrophy (Charcot Marie-Tooth disease)

WITH SENSORY CHANGES
Lesions of nerve plexuses or peripheral nerves
Nerve root or spinal cord compression

ATROPHY WITHOUT SENSORY CHANGES

The several disorders in this group are distinguished as usual by their characteristic clinical features. There are, however one or two obvious characteristics that at once narrow down the possibilities.

The hereditary disorders are the muscular dystrophies, dystrophia myotonica and peroneal muscular atrophy the family history and the early age of onset exclude motor neurone disease. Again, fibrillation of wasting muscles is very obvious only in motor neurone disease it is absent in the muscular dystrophies.

Arthritic muscular atrophy

This condition affects principally the extensor muscles of the joint. The degree of wasting varies with the type of joint disease likely to be very marked in joint tuberculosis it may be almost as severe in rheumatoid arthritis but as a rule is less so in osteo-arthritis.

Disuse atrophy

Disuse atrophy follows prolonged splinting, or inactivity of part of a limb from any cause. It is a common finding in the muscles of the legs when activities are reduced by arteriosclerosis obliterans.

The residual paralysis of poliomyelitis might be confused with the muscular dystrophies but the clinical course of each is strikingly different. Soon after the febrile onset of acute poliomyelitis, paralysis

appears suddenly reaching its maximum intensity in the first few hours. After a period of partial recovery residual paralysis is stationary—in marked contrast to the slowly progressive course of a muscular dystrophy Some muscles remain totally paralysed, and waste rapidly and completely others waste, but are capable of some degree of recovery The residual paralysis is asymmetrical

The muscular dystrophies

These are familial diseases of which there are three clinical types. Common to all are progressive symmetrical weakness of shoulder and hip girdle muscles, winged scapula and lordosis.

Pseudohypertrophic Type In this variety boys are affected almost exclusively Symptoms appear when he begins to walk he falls easily and gets up with difficulty from weakness of the muscles of the pelvic girdle The type owes its name to enlargement of some of the wasted muscles from increase in the connective tissues, notably enlargement of the calf muscles, the glutei the spinati, the erector spinae. Other muscles are obviously wasted from the first, and the apparently enlarged muscles shrink later Lordosis compensates for a tilting forwards of the pelvis from weakness of the glutei—the extensors of the hip Ultimately all the trunk and limb muscles are affected Most patients die within 10 years.

Scapulohumeral Type This variety begins in the upper arm and shoulder winged scapula may be the first sign appearing in adolescence.

Facioscapulohumeral Type This variety often begins in early childhood affecting the facial muscles first.

Myotonia atrophica (DYSTROPHIA MYOTONICA)

This is a hereditary muscular dystrophy with myotonia—a persistence of muscular contraction after voluntary or electrical stimulation.

The onset is usually between the ages 20-30 and it occurs in men more often than in women Muscular wasting slowly progresses over some years, affecting the distal muscles of the limbs, and characteristically the muscles of the face and the sternomastoids. Myotonia slows up movements It can be well seen in the slow relaxation of the fingers after gripping. Some wasted muscles may not show this phenomenon. The affected muscles do not show fibrillation. There are manifestations of the hereditary defect in other systems notably cataract, and impotence from testicular atrophy

Peroneal muscular atrophy (CHARCOT–MARIE–TOOTH DISEASE)

This is an uncommon muscular atrophy resembling the muscular

dystrophies in being a familial disease, and *beginning in childhood It is* not a primary disease of muscle, but is due to degeneration of motor neurones.

The age of onset is 5-10 years, but it has been known to begin at any time in the first half of life Sensation may be normal, but there may be some impairment over the affected limbs. Weakness and wasting begin in the small muscles of the feet, the peronei and the extensors of the toes. The result is foot drop, pes cavus, and claw toes. Atrophy spreads slowly usually over a period of many years, up the limb stopping when it has involved the lower third of the thigh, and leaving the so-called champagne bottle thighs.

Some years after the onset, the small muscles of the hand are affected, muscular wasting spreading slowly up the forearm to the elbow There is sometimes fibrillation of the wasting muscles.

The combination of claw foot and claw hand in an adolescent is pathognomonic.

Motor neurone disease

Progressive Muscular Atrophy This is a clinical variant of motor neurone disease, in which the anterior horn cells of the cervical region are chiefly affected It affects typically the middle-aged man. His first complaint is of wasting of one hand first of the thenar eminence then the interossei, finally all the small muscles of the hand Wasting spreads up the limb as a rule very slowly being confined to the hand and fore arm for several years. Finally the whole of the limb is affected. Perhaps about a year after the onset, atrophy starts in the opposite hand, and is followed by the same progress up the limb

The condition is painless, except for some mild aching in the affected arm and slight paraesthesia. Sensation is normal. Fibrillation of the wasting muscles is marked moreover it precedes atrophy its presence in the apparently healthy muscle of the arms being commonly observed, and being a sign that atrophy will soon appear Winging of the scapula is very uncommon The tendon jerks in the arms may be diminished normal, or sometimes increased.

Amyotrophic Lateral Sclerosis In this clinical variety of motor neurone disease the chief impact of the neurone degeneration is on pyramidal neurones. It differs from progressive muscular atrophy only in the presence of pyramidal signs and of a moderate weakness of the legs. Tendon jerks in both upper and lower limbs are brisk there is ankle clonus, but an extensor plantar response is very late in appearing.

The chief points distinguishing progressive muscular atrophy and the muscular dystrophies are shown in the table

	MUSCULAR DYSTROPHY	PROGRESSIVE MUSCULAR ATROPHY
Hereditary factor	present	absent
Age at onset	early childhood or adolescence	middle age
Muscles first affected	limb girdles	hands
Symmetry	symmetrical	asymmetrical
Fibrillation	absent	marked

ATROPHY WITH SENSORY CHANGES

Having found that atrophy is, or has been accompanied by sensory symptoms, or that sensory loss can be demonstrated we now want to know if we are dealing with a peripheral nerve or nerve plexus lesion or with nerve root or spinal cord compression An important clue is to be found by observing which muscles are included in the selective wasting. The muscle distribution of a peripheral nerve is well known and is different from that of any spinal segment or segments. Some important instances are shown in the tables.

TABLE SHOWING THE DISTRIBUTION OF THE SEVERAL NERVES OF THE UPPER AND LOWER LIMB

NERVE	ROOT ORIGIN	MUSCLES
Axillary	C.5 C.6	Deltoid
Long thoracic	C.5 C.6, C.7	Serratus anterior
Musculocutaneous	C.5 C.6, (C.7)	Biceps, coracobrachialis, brachialis
Radial	C.5 C.6, C.7 C.8, T 1	Triceps, extensors of fingers and wrist, brachioradialis, supinator
Median	C.5, C.6, C.7 C.8 T 1	Flexor carpi radialis and digitorum sublimis, lateral half flexor digitorum profundus, pronators, opponens and abductor pollicis, 2 lateral lumbricals
Ulnar	C.7 C.8, T 1	Flexor carpi ulnaris, medial half flexor digitorum profundus, interossei, 2 medial lumbricals, hypothenar muscles, adductors of the thumb
Obturator	L.2, L.3 L.4	Adductors of thigh
Femoral	L.1 L.2, L.3 L.4	Quadriceps, iliopsoas, sartorius
Tibial	L.4 L.5 S 1 S.2, S.3	Calf muscles, tibialis posterior flexor digitorum and hallucis longus, intrinsic muscles of the foot
Common peroneal	L.4 L.5 S.1 S.2	Peronei, tibialis anterior extensor digitorum longus and extensor hallucis longus

Lesions of individual peripheral nerves

Radial Paralysis The nerve may be injured as it passes round the humerus the triceps escapes Or it may be injured in the axilla from pressure of a crutch and the triceps is involved The diagnostic signs are wrist drop and loss of supination. Sensory loss is slight and inconstant it may be found on the dorsum of the first and second metacarpals.

SEGMENTAL INNERVATION OF THE MUSCLES OF THE UPPER EXTREMITIES (simplified)

C.4 C.5 C.6 C.7 C.8 T 1

supraspinatus
infraspinatus
deltoid
biceps
brachialis
triceps
supinator
brachioradialis
ext carpi rad.
extensors of the digits
flexor carpi rad.
flexor carpi uln.
ext. carpi uln.
flexors of the digits
small
hand
muscles
lumbricals
interossei

Ulnar Paralysis. The nerve may be injured behind the medial epicondyle of the humerus, either by trauma, or by compression by osteophytes in osteo-arthritis of the elbow Wasting is obvious in the hypothenar muscles and the interossei. The diagnostic signs are

1 *Weak flexion of ring and little fingers and of the wrist* on attempted flexion the wrist is drawn to the radial side from weakness of the flexor carpi ulnaris.

2 *Inability to spread and approximate the fingers* from paralysis of the interossei.

3 *Claw-hand position of ring and little fingers*—e.g. hyperextension at the metacarpo-phalangeal joint, flexion at the interphalangeal joints. (The interossei with the lumbricals, flex the first phalanges and extend the second and third.)

4 *Paralysis of the adductors of the thumb* is seen in Froment s thumb sign. A piece of paper is held by finger and thumb of each hand. On the

SEGMENTAL INNERVATION OF THE MUSCLES OF THE LOWER EXTREMITIES (simplified)

L.2 L.3 L.4 L.5 S.1 S.2

gluteus medius
gluteus minimus
gluteus maximus
adductor longus
adductor brevis
adductor magnus
adductor minimus
quadriceps femoris
semitendinosus
semimembranosus
biceps femoris
tibialis ant.
ext. digitorum long.
ext. hallucis long
soleus
gastrocnemius
peroneus
tibialis posterior
flexor hallucis longus
flexor digitorum longus
small
foot
muscles
lumbricals
interossei

paralysed side this can be achieved only by flexing the terminal phalanx of the thumb

5 *Sensory loss* in a complete ulnar paralysis is found on the ulnar margin of the hand, and over the little finger and ulnar side of the ring finger

Median Paralysis This is uncommon. The nerve may be compressed in the carpal tunnel, when the hand alone is affected. Wasting shows as a hollow in the radial half of the thenar eminence. The thumb cannot be opposed or flexed, and in long-standing cases tends to fall into the plane of the other digits with the nail facing dorsally (*main de singe*) Flexion of the index and middle fingers is impaired but the ring and little fingers flex normally *Sensory loss may be found on the radial half*

of the palm lateral to the prolongation of a line down the middle of the ring finger

When the nerve is injured in the upper arm, the flexor carpi radialis, part of the flexors of the fingers the flexor pollicis longus, and the pronator are also affected. The power of pronation is lost Flexion of the wrist is weak, and the wrist is pulled over to the ulnar side on attempted flexion.

Multiple peripheral neuritis

This is a bilateral affection of the distal parts of peripheral nerves. There are several clinical types, but possibly a single toxic metabolite is the responsible agent in most of them. The main clinical features are

1 Tingling in hands and feet.

2. Impaired sensation in glove and stocking areas.

3 Atrophic paralysis affecting the lower limbs earlier and more severely than the upper and the *distal* muscles more severely than the proximal. In the legs the anterior tibial and peroneal muscles are affected causing foot drop in the arms the brunt falls on the extensors of the wrist and fingers, causing wrist drop.

4 The affected muscles are tender painful and subject to cramps.

5 There is hyperaesthesia of the soles.

There is no essential variation of these clinical features with the cause of the neuropathy The main causes are

EXTRINSIC POISONS

Alcohol many metals many organic substances e.g. carbon bisulphide apiol (used as an abortifacient) etc.

INTRINSIC POISONS

Diabetes bronchial carcinoma polyarteritis nodosa.

VITAMIN DEFICIENCY

Beriberi pellagra.

INFECTIONS

Acute febrile polyneuritis diphtheria leprosy

Alcoholic Neuritis. The earliest complaint is of tingling, numbness and pain in the feet, aching, tired legs and night cramps in the calves. The calf muscles are tender The legs slowly become weaker with increasing foot drop. In long-standing cases wasting may be severe. The ankle jerks are lost. There is increasing sensory loss beginning over the feet and spreading proximally perhaps ultimately causing ataxia. Less often, atrophic paralysis affects the extensors of the wrist, causing wrist drop.

Diphtheria A very similar multiple peripheral neuritis may occur in diphtheria. Recovery comes in about 6 months.

Diabetic Neuritis There has been in recent years a renewed interest in diabetic neuropathy Usually peripheral nerves are affected sometimes the spinal cord It is said to occur in more than 50 per cent. of diabetic patients both in hyperglycaemia when symptoms respond quickly to diabetic control and in controlled diabetes, when spontaneous remissions and relapses are very common.

The symptoms are paraesthesia and pain in the legs, tender calf and plantar muscles, variable muscle weakness and wasting, sensory loss especially loss of vibration sense, absent or diminished ankle and knee jerks.

Pain is the commonest symptom worse at night and often described as cramp in the legs, or severe burning pain in the feet. It is sometimes unilateral Cases have been reported of severe muscular wasting affecting the quadriceps, or all the muscles of one leg.

Bronchial Carcinoma The existence of a causal relation between bronchial carcinoma and peripheral neuropathy is now accepted A purely sensory polyneuritis, and the classical mixed type have been described. Usually the presenting syndrome is a polyneuritis the carcinoma is discovered on investigation.

Beriberi This is caused by the absence of thiamine (vitamin B_1) in the diet, and is rarely seen in this country It is a severe multiple peripheral neuritis with atrophic paralysis and oedema of the legs.

Lead Palsy This has usually been grouped with peripheral neuritis, but it is now known to be a myopathy due to a direct toxic action of lead on the muscles. The pathological change is a degeneration of anterior horn cells, motor nerve fibres and muscle fibres. The symptoms are purely motor Bilateral wrist drop, with no pain and no complaint of paraesthesia, is characteristic. Other symptoms and signs of plumbism—nausea, vomiting, epigastric pain, colic, blue line on the gums, anaemia with punctate basophilia—establish the diagnosis.

Acute Febrile Polyneuritis This is a disease of unknown causation, possibly a virus infection, liable to be confused with acute poliomyelitis. There is complaint of severe numbness and tingling in the extremities and a rapidly spreading symmetrical paralysis in arms and legs, includ ing proximal limb muscles, and perhaps abdominal and respiratory muscles. If the patient does not die from respiratory failure, recovery is equally rapid When the paralysis has attained its full extent, it begins to abate and within 2 or 3 weeks recovery is complete

Nerve root or spinal cord compression

Most instances of muscular atrophy from nerve root or cord

compression present as pain they are discussed under pain in the arm [p. 73], and pain in the leg [p. 171]

There are two groups of signs of cord compression [p. 213]

SEGMENTAL SIGNS

Sensory root pain (which may precede by many months symptoms of cord compression)

Motor wasting of muscles innervated by the affected spinal segment.

SIGNS FROM INTERFERENCE WITH TRACTS IN THE CORD

Symptoms below the level of compression develop as this increases in severity At first asymmetrical, if the compression is asymmetrical, they comprise

1 pyramidal signs in the homolateral leg
2. impairment of pain and temperature sense in the contralateral leg.

Syringomyelia

Syringomyelia is distinguished from other causes of cord compression by among other characters, its *painless* nature.

It is a gliosis and cavitation developing in the region of the grey commissure, and extending in the length of the cord over a number of segments. It is most advanced in the cervical segments.

Symptoms begin in adolescence or early adult life.

Wasting first appears in the intrinsic muscles of the hand, affecting all of them. It may be unilateral for a long time, or at least much more marked on one side than the other but it gradually becomes symmetrical. It then spreads in regular fashion to the forearm, where the flexors are chiefly affected the upper arm, the sterno-mastoid, trapezius, and other muscles of the neck and shoulder girdle.

Trophic changes appear in the hands the skin becomes cold and livid and perhaps abnormally dry and *a hard oedema develops on the* dorsum. Ultimately the hand has an atrophic glossy skin.

Various *deformities of the hand* are a sequel sometimes claw hand sometimes thumb and index partly flexed, with the other fingers in complete flexion contracture sometimes wrist and fingers in hyper extension (*main de predicateur*)

In addition superficial ulcers and perhaps painless whitlows develop on the hands, for the skin has lost pain and temperature sense. Over a sleeved jacket area (the jacket usually not extending lower than the 6th rib) there is *dissociated anaesthesia* not a loss of all forms of sensation, but only of pain and temperature sense touch and postural sense

remain normal Over this area the patient cannot distinguish heat from cold, nor a pin-prick from touch He does not feel burns or other injuries, and to this sensory loss are due the painless injuries on the hands so characteristic of the disease and so unaccountable to the patient.

In one lower limb it is not rare to find pyramidal signs increased knee and ankle jerks, and extensor plantar response and a slight degree of spasticity

Syringomyelia may be confused with progressive muscular atrophy But fibrillation in the wasting muscles is much less marked than in this condition. Also in contrast to progressive muscular atrophy are the trophic changes in the hands, and the dissociated anaesthesia.

ATROPHY OF INTRINSIC MUSCLES OF THE HAND

Causes

WITHOUT SENSORY CHANGES

Arthritic muscular atrophy
Peroneal muscular atrophy (late stage) [p 19]
Progressive muscular atrophy [p 10]
Amyotrophic lateral sclerosis [p 10]

WITH SENSORY CHANGES

Ulnar paralysis [p 12]
Median paralysis [p 13]
Cervical rib syndrome [p 81]
Syringomyelia [p 16]
Nerve root and spinal cord compression [p 15]
disease of vertebrae affecting T 1 segment Pancoast tumour pachymeningitis spinal cord tumour
Lower brachial plexus injury

Progressive muscular atrophy begins in the outer half of the thenar eminence. It might therefore be confused with median paralysis, or the median type of cervical rib syndrome. If first seen when it has spread to all the intrinsic muscles, the condition most closely resembling it would be syringomyelia, which from the outset affects all the hand muscles.

It is important therefore to know if the condition is painful or not. The two observations most helpful diagnostically are 1 sensory loss on the affected hand or elsewhere on the upper limbs 2. fibrillation in the wasted muscles.

In progressive muscular atrophy sensory phenomena are absent, and the wasted muscles show marked fibrillation—as may apparently healthy muscles in the arm In median paralysis the cervical rib syndrome, and syringomyelia, fibrillation is inconstant, slight or absent.

The only similar condition in which fibrillation is well marked is peroneal muscular atrophy [p 9] but this is a familial disease beginning in childhood and first affecting the feet and legs there is marked wasting of the leg muscles before the hands are affected.

Other conditions causing wasting of the small muscles of the hand are

1 Ulnar paralysis [p. 12] which affects the hypothenar muscles the interossei, and the two inner lumbricals the signs are very characteristic

2. A lesion compressing T 1 nerve root or the spinal cord at this level The association of severe root pain will suggest the diagnosis.

3 Lower brachial plexus injury This is uncommon involving C.8 and T 1 roots, it causes atrophic paralysis of the intrinsics of the hand, and the flexors of the wrist and fingers.

ATROPHY OF MUSCLES OF THE LOWER LIMB

The responsible conditions are discussed in the general section on muscular atrophy and under pain in the leg [p 171] The more important causes may be summarized here

WITHOUT SENSORY CHANGES

Arthritis knee—quadriceps hip—glutei.

Disuse from joint disease from arteriosclerosis.

Poliomyelitis

Muscular dystrophies pseudo-hypertrophic type.

Peroneal muscular atrophy

WITH SENSORY CHANGES

Multiple peripheral neuritis [p. 14]

Sciatic neuralgia [p. 161] due to

Cauda equina compression by meningiomata or other tumours or gross disc protrusion

Nerve root compression disc protrusion osteo-arthritis disease of the lumbar spine.

Trauma of the sciatic nerve

Single nerve palsies

Femoral

Obturator

Common peroneal

Tibial

Single nerve palsies in the lower limb are comparatively uncommon

Compression of the femoral nerve by intra abdominal growth, or during parturition, is uncommon It affects the quadriceps and ilio-psoas, causing weakness of flexion at the hip and extension at the knee and loss or diminution of the knee jerk A femoral neuralgia may also be due to serious disease of the spine affecting lumbar roots L.2, 3 4

Injury to the obturator nerve is followed by inability to adduct the thigh the patient cannot cross the knee of the affected leg over the other

The common peroneal nerve [p 53] is subject to injury as it winds round the fibula and to a neuritis of unknown origin. It causes a foot drop from weakness and wasting of the peronei tibialis anterior and extensors of the toes.

The tibial nerve is much less commonly injured the result is a paralysis of the calf muscles.

ATROPHY OF SHOULDER GIRDLE MUSCLES

There is usually no difficulty in recognizing arthritic muscular atrophy disuse atrophy or the residual paralysis of poliomyelitis Apart from these, the causes may be grouped

WITHOUT SENSORY CHANGES

Muscular dystrophies [p 9]

Paralysis of serratus anterior (winged scapula)

WITH SENSORY CHANGES

Single nerve injuries supra-scapular—deltoid axillary—supra spinatus and infraspinatus.

Upper plexus injuries [C.5 6] deltoid, supraspinatus and infra spinatus, serratus anterior rhomboids, clavicular part of pectoralis major (and some arm muscles)

Nerve root and cord lesions [p 15]

C.4—trapezius, levator scapulae

C.5—deltoid supraspinatus and infraspinatus, rhomboids.

Radiculitis [p 174]

The muscular dystrophies are unlikely to be confused with any condition in the second group They are essentially familial diseases characterized by a progressive symmetrical weakness of shoulder and hip girdle muscles, beginning in early childhood or adolescence.

Instances of atrophic paralysis following injury also present no diagnostic problem.

Winged scapula

Paralysis of the serratus anterior causes the vertebral border of the scapula to stand away from the chest wall when the arm is raised forwards at the shoulder to a horizontal position—and particularly when in this position it is thrust forwards against resistance The patient s complaint is of a general weakness of the arm and inability to elevate it fully

The nerve supply to the muscle is the purely motor long thoracic nerve (C.5 6 and 7) Where it descends on the anterior surface of the scalenus medius, it is liable to injury from undue pressure on the shoulder, as in carrying an army pack.

Possibly some of these cases are due to a brachial radiculitis [p. 174] in which it may be that long thoracic fibres are mainly affected. It would seem also that the nerve may be affected by a neuritis after various infective diseases.

Winged scapula is fairly commonly noticed in the cervical rib syndrome [p. 81] in which presumably the long thoracic nerve is stretched.

Finally the serratus anterior is one of the first muscles to be affected in the muscular dystrophies [p 9]

BONE, SWELLINGS OF

Bone swellings, discovered clinically or radiologically may be grouped as due to

TRAUMA

INVASIVE DISEASES

Infections pyogenic tuberculous syphilitic.

New growths

primary malignant tumours osteogenic sarcoma fibrosarcoma Ewing s tumour giant cell tumour

metastases

benign tumours osteochondroma enchondroma osteoid osteoma.

BONE CYSTS

BONE DYSTROPHIES Paget s disease acromegaly

Localized swelling after injury to a bone—from effusion under the periosteum, which after subsidence may leave a permanent thickening.

or from callus after a fracture—usually presents no diagnostic difficulty but there are times when an ossifying haematoma closely mimics a sarcoma.

INFECTIONS

An acutely painful, tender swelling near the end of a long bone, with red and oedematous overlying skin, may be due to an acute pyogenic periostitis and osteomyelitis [p 157]

A chronic staphylococcal bone abscess is called Brodie s abscess. It occurs most often near *one end of a long bone, usually the tibia, though* it may be femur or humerus. Dense sclerosed bone walls in the abscess, and with periosteal thickening may bring some enlargement of the bone end as a late event.

Rarely a chronic tuberculous abscess occurs in young adults, near one end of a long bone, particularly the upper end of the tibia. It may easily be confused with Brodie s abscess there is the same aching pain, worse at night, with local tenderness swelling may be considerable. Tuberculous dactylitis in children is an osteitis of a phalanx, metacarpal, or metatarsal. The bone is expanded new periosteal bone keeping pace with destruction within.

Syphilis of bone is very rare in this country There are two forms syphilitic periostitis, and endosteal gumma.

Periosteal nodes may occur on the long bones, or a diffuse periostitis with sclerosis of the bone beneath. This in the tibia gives a fusiform swelling replacing the sharp anterior crest of the bone.

Gummatous osteitis affects most commonly the clavicle or the tibia the skin overlying the swelling may ulcerate An endosteal gumma is usually in the cancellous end of a long bone, which is expanded both by the granuloma and new periosteal bone It may be mistaken for a malignant growth.

NEW GROWTHS

Primary malignant tumours

Osteogenic Sarcoma. This neoplasm affects mainly children, adolescents and young adults. It is very rare after 40. It may follow trauma but whether this merely calls attention to the condition or is causally connected is unknown.

It occurs usually at the end of a long bone, and most commonly near the knee. The first symptom is pain—increasingly severe continual pain, disturbing sleep. It is thought to be due to stretching of the periosteum and is followed after a few weeks by swelling. The swelling may become a large fusiform tumour and when near the knee be increased in bulk

by effusion into the joint It is firm and elastic, or soft fluctuation is very rare.

The diagnosis cannot be made on the clinical features. It can often, but not always, be made with the additional data provided by X ray films of the tender bone end But many instances do not give a charac teristic appearance, and films may have to be repeated at frequent

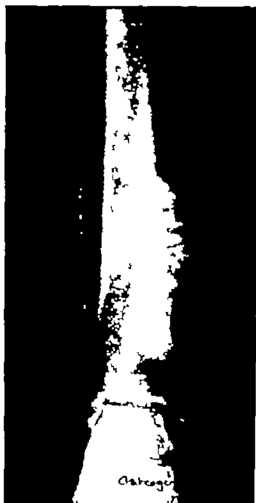


FIG. 2a
Osteogenic sarcoma classical sun-ray appearance.

intervals. There are some who object to biopsy on the ground that there is a risk of dissemination but the general tendency now is to regard it as safe and useful. Even the histological findings, however are at times equivocal with all available data mistakes are still possible.

X ray appearances may be typical and diagnostic fine sharp spicules of bone set on the shaft at right angles to it longest at the site of the tumour diminishing peripherally the bone beneath may be increased in density [FIG 2*a*] But in many instances X ray appearances may be confused with those of syphilitic or tuberculous osteitis, chronic osteomyelitis, or ossification of a haematoma In the osteolytic type of



FIG 2*b*

Osteogenic sarcoma osteolytic type
(by kind permission of Dr F Campbell Golding).

growth the tumour may be visible in the soft parts and here the cortex is seen eroded, with variable destruction of the underlying bone. There is no new bone formation the appearance of bone destruction fading imperceptibly into that of normal bone. Pathological fracture is common in osteolytic growths [FIG 2*b*] In the sclerosing, or osteoplastic, type the affected bone is densely sclerosed, its outline is blurred or eroded, and from it project irregular bosses and spicules of new bone.

When bone sarcoma is suspected, the chest should also be X rayed,

Metastases may be present in the lungs at an early stage their detection will save a useless operation.

Histologically these tumours show considerable variation. The tumour cells—spindle shaped round, or polygonal—are in a stroma of varying amount, and composed of myxomatous, cartilaginous, osteoid, or bony tissue in very varying proportions.

Fibrosarcoma. This is a rare tumour arising from the periosteum, or less commonly within the bone. In most instances the periosteal variety is found near the knee causing a firm and smooth swelling over the



FIG. 3
Ewing's tumour
(by kind permission of Dr F Campbell Golding).

upper end of the tibia, or lower end of the femur X ray films show the soft tissue shadow and erosion of the bone cortex in contact with it.

Ewing's Tumour This tumour is also very rare it was formerly called round cell sarcoma, or endothelioma of bone.

Most instances are in young people, usually 15-25 years. It usually occurs in the shaft of a long bone, most often tibia, fibula, or femur but it may be in any bone The first symptoms are recurrent attacks of pain and pyrexia, suggesting chronic osteomyelitis. After some months

of this a tender swelling of the bone appears. Metastases occur in the lymph glands, and especially in other bones, notably the skull

The X ray appearances are of destruction of cancellous bone [FIG 3] In the shaft of a long bone when the tumour reaches the surface plaques of new periosteal bone are laid down over it in onion like layers the innermost layers are continually absorbed by the expanding tumour and new layers are deposited on the outside. Periosteal proliferation increases the local density and pathological fracture is rare

The tumour is extremely radio-sensitive, and the response to X radiation may be used as a therapeutic diagnostic test.

Giant Cell Tumour (Osteoclastoma) This tumour is only locally malignant. The age incidence is 16-25 years. It occurs most commonly above or below the knee in the epiphyseal region

The first complaint is of a swelling it is accompanied by pain though less severe than in sarcoma. A pathological fracture occurs in about 14 per cent. of cases.

The X ray appearances of the affected part are of an expanded bone with the cortex reduced to a thin shell. Coarse trabeculae crossing the tissue give it a characteristic soap bubble appearance [FIG 4a]

Metastases

Secondary deposits of carcinoma are rare in the limb bones, except at the upper end of the femur and humerus they are commonest in vertebrae [p 93], pelvis, rib sternum, and skull—bones containing red marrow

The prominent symptoms are pain and local tenderness if there is swelling it is unlikely to be detected They are most often osteolytic, and pathological fractures occur in about 15 per cent.

Benign tumours

Ecchondroma. This is a cartilage tumour projecting from a bone. It later ossifies, becoming what is called an exostosis, or osteochondroma. It is common near the knee, and may occur at the upper end of the femur It may grow to such a size as to interfere with the mechanics of the joint.

Enchondroma. This is a cartilage tumour within a bone, expending the bone locally reducing the cortex to a thin shell. Commonest in the metacarpals, and phalanges of the fingers, they may be found near the end of a long bone, single or multiple. Swelling is the only clinical sign it is not painful. But after many years it can become malignant, and then it becomes painful, rapidly increases in size and breaks through the cortex.

Osteoid Osteoma This benign tumour first described in 1935 occurs mainly (75 per cent.) in adolescents and young adults It is rare after 30, but a few after 50 have been recorded A single tumour it is commonest in the lower limb, in the cancellous tissue or the cortex of a long bone or a phalanx. The prominent symptom is localized persistent pain, worse at night, and local tenderness. There may be swelling of the adjacent soft tissues. Near a joint it may cause a synovitis.

The tumour is composed of irregularly arranged areas of osteoid, in a vascular matrix



FIG. 4a

Osteoclastoma
(by kind permission of Dr F Campbell Golding)

X ray films show a rounded clearly demarcated area, usually less than 2 cm. in diameter at first translucent. Later when osteoid calcifies, it becomes radiopaque. But the tumour may be hidden in dense periosteal new bone [FIG 4b]

BONE CYSTS

A bone cyst radiologically is a rounded and fairly sharply demarcated area of translucency The commoner conditions responsible are

1 Cysts following trauma

2. Cysts in the metacarpals or phalanges due to degeneration of enchondromata



FIG. 46

Osteoid osteoma
(by kind permission of Mr W D Coltart)

3 Tumours of bone, e g. enchondroma osteoclastoma

4 Chronic infective conditions e.g. Brodie s abscess [p 21]

5 Solitary cysts near the extremity of femur humerus, or tibia, of unknown nature. They are associated with swelling and aching, and are sometimes the cause of spontaneous fracture.

BONE DYSTROPHIES

Paget's disease

This is a bone dystrophy beginning in middle age or later affecting perhaps a single bone more often many but never the whole skeleton. Most commonly affected in order of decreasing frequency are sacrum, skull, lumbar vertebrae, pelvic bones, bones of the lower limbs.

It is very slowly progressive the affected bone becomes enlarged and at first softer than normal. Swelling may be obvious in the tibia and a need for larger hats each year calls attention to a progressive enlargement of the skull In one of Sir James Paget's cases, the circumference of the skull increased by 5 inches in 30 years. Softening of the bone brings deformities a marked anterior curvature of tibia or femur a slowly increasing kyphosis sometimes compression of a vertebral body But gross deformities are rare.

In the later years of the disease the bone becomes heavier and denser than normal In this stage of increasing sclerosis no further deformity will occur

Sometimes the condition is painless, and only discovered during X ray examination for some other condition. Usually however the affected bones are a source of continual deep aching pain, which may at times be quite severe. X ray examination, carried out either because of pain, or because of a spontaneous fracture, establishes the diagnosis the appearances are quite characteristic.

The primary lesion in Paget's disease is bone destruction. This is countered by erratic repair a very marked overgrowth of bone, but haphazard, not following the normal trabecular architecture. The two processes go on simultaneously in the skull it is sometimes possible to see an area of resorption and an area of repair side by side. The result is that normal bone is replaced by coarse trabecular masses and a greatly thickened cortex. The other essential change is replacement of the marrow by a cellular fibrous tissue of extreme vascularity The blood flow in Paget bones has been found to be increased up to 20 times the normal.

The bone destruction is not an exaggeration of the normal process of bone resorption, as in hyperparathyroidism. Serum calcium and phosphorus are normal. The very marked osteoblastic activity responsible for the overgrowth of bone is reflected in a very high serum alkaline phosphatase level

X-ray appearances are usually unmistakable [FIG 5a]. The essential changes are 1 enlargement of the bone 2 replacement of normal bone structure by a coarse haphazard trabeculation 3 at the edge of a zone of overgrowth may be found normal bone and small zones of rare-

thick, the medullary canal narrowed The normal bone structure has been replaced by coarse spongy bone of varying density

Acromegaly

Hyperpituitarism after epiphyseal growth has ceased causes the increase in thickness of bones called acromegaly The arresting signs are enlargement of hands and feet, jaws, nose, tongue, lips and ears.



FIG 5b

Paget's disease skull
(by kind permission of Dr G A. S. Lloyd).

A man, or a woman—the sexes are affected almost equally—aged 20-40 years, begins to notice this alteration in the features, and in the size of the hands and feet. The fingers become clubbed, the hands broad and spade like. There is complaint of intense lassitude and muscular weakness, and of pains in muscles, bones and joints. The heart is usually enlarged, as are all internal organs. Arteriosclerosis is a constant development. Sexual functions are depressed amenorrhoea and virilism appear in women.

When the hormone excess is from a pituitary tumour headache may be a prominent complaint, and bitemporal hemianopia may develop. X ray films of the skull may show enlargement of the sella—but not always Films of the limb bones frequently show exostoses at the site of muscle insertions.

BONE TENDERNESS

Local tenderness occurs whenever periosteum is 1 inflamed either by trauma or infection e g repeated blows on a bone near the surface as in the tender shins of footballers or in osteomyelitis 2 stretched by an expanding lesion, e g subperiosteal extravasation of blood in scurvy or primary new growths

General tenderness is a feature of the generalized rarefying diseases of bone.

RAREFYING DISEASES OF BONE

Bone rarefaction is an X ray finding It may affect the whole skeleton or only parts of it. The metabolic diseases of bone are generalized but some of the local or disseminated disorders not affecting the whole skeleton, may closely resemble them radiologically and therefore come into the differential diagnosis.

The presenting symptom in the metabolic diseases of bone is likely to be pain in the back and limbs and is often ascribed to rheumatism Palpable bones are tender

The rarefying diseases of bone may be grouped as follows

METABOLIC DISEASES

Inadequate Deposition

failure of calcification osteomalacia

osteoblastic failure osteoporosis

Excessive Resorption

Hyperparathyroid osteitis fibrosa generalisata

LOCALIZED

Invasive diseases myelomatosis, carcinomatosis skeletal xanthomatosis.

bone dystrophies polyostotic fibrous displasia

METABOLIC DISEASES OF BONE

The skeleton as a mechanical structure is of secondary importance to the skeleton as a calcium depot The ionized calcium of interstitial fluid is one of the several constants of the body essential for cell life. Just as to maintain a constant glucose level the animal has a reserve of glycogen in the liver so it depends for the maintenance of its Ca^{++} constant on the immense reserves of the skeleton Calcium ions are continually removed from the skeleton to provide the body's needs, and continually

replaced by ingested calcium. A balanced decalcification and re-calcification proceeds without cessation throughout life. But it is not a simple matter of solution and precipitation decalcification is accompanied by *resorption of the organic matrix, and re-calcification by the* deposition of new bone matrix. The balance is really one of bone resorption and deposition.

The metabolic diseases of bone result from a disturbance of this balance either resorption is excessive (hyperparathyroidism) or deposition inadequate (osteomalacia osteoporosis)

Bone resorption is controlled by the parathyroid hormone, and a fall in serum calcium is the normal stimulus to its secretion.

Bone deposition is a complex process. Osteoblasts first lay down the osteoid tissue which is to be calcified. At the same time these cells produce the enzyme alkaline phosphatase, which determines the deposition in this matrix of a calcium phosphate carbonate complex called dahlite.

Fluid bathing the matrix contains calcium and phosphate ions in almost saturated solution an increase in phosphate ions determines prompt precipitation of calcium phosphate. The action of alkaline phosphatase is to liberate phosphate ions from non ionized organic phosphate compounds. The level of serum alkaline phosphatase is a measure of osteoblastic activity

Biochemical Tests. Those of most value in distinguishing the several metabolic diseases of bone are the estimations of serum calcium, phosphate, and alkaline phosphatase.

Serum calcium is normally 9-11 mg. per 100 ml.

Serum inorganic phosphate is 5-6 mg. per 100 ml. in infancy gradually falling to 3-4 5 mg. per 100 ml. in adults.

Serum alkaline phosphatase, measured in King Armstrong units, is 3-13 units per 100 ml. in normal adults 10-30 units in normal children. It is highest in the first 2 years of life, and gradually falls to the adult level.

X ray Appearances In all three types there are 1 excessive radio-translucency 2. if the intervertebral discs have not lost all turgidity through the degenerative changes of age, they bulge into the softened bone, becoming markedly bi-convex, and the bodies bi-concave 'fish vertebrae 3 crushing and wedging of one or more vertebral bodies.

In osteomalacia and in hyperparathyroidism there are additional characteristic features that will be mentioned in the appropriate place.

Osteomalacia

Osteomalacia is a rarefaction of the whole skeleton from inadequate

bone deposition due to a failure of calcification. Osteoid—the organic matrix—is laid down normally but is not calcified Osteoblastic activity is in fact increased the error is a lack of available calcium.

The serum levels of calcium and phosphate depend in part on the degree of compensatory parathyroid secretion The usual finding is normal calcium and low phosphate but it may be low calcium and normal phosphate, or both may be low What determines failure of calcification is a low calcium and phosphate product. Calcium and phosphate ions in a body fluid are in a state of equilibrium such that a rise in calcium ions leads to a fall in phosphate ions, and vice versa. The product of serum calcium and serum phosphate, expressed in mg. per 100 ml., is constant it is 30-40 for adults 40-55 for growing children. Rickets occurs in children if the product is below 35

The serum alkaline phosphatase is increased.

These biochemical changes may be compared at once with those found in osteoporosis, and in osteitis fibrosa generalisata.

	OSTEOMALACIA			OSTEOPOROSIS	OSTEITIS FIBROSA GENERALISATA
Serum Ca	normal	low	low	normal	high
Serum P	low	normal	low	normal	low
Serum alk. phosphatase	high			normal	high

The cause of the lack of available calcium in osteomalacia is usually a deficiency of vitamin D

Rickets. This disease results from the effect of a simple deficiency of vitamin D on growing bones. It is osteomalacia with certain very characteristic changes in the growing ends of the long bones. It is readily curable by the administration of small doses of vitamin D

With improved infant feeding, rickets has become a rarity in this country It begins in infancy (6 months to 2 years) At the growing ends of the bones all normal regularity in the process of bone growth is absent. The epiphyseal discs are enlarged by over production of osteoid, causing the characteristic drum-stick swellings of the bone ends. Over production of osteoid leads also to bossing of the skull, and enlargement of the costochondral junctions ('rickety rosary')

A moderate dwarfism comes from old rickets, from stunting of the lower limb bones, and increase in their normal curvature possibly with a contribution from kyphosis and scoliosis. From these stunted bent bones will be found either genu valgum or genu varum a big square skull with a bulging forehead a deformed thorax—rickety rosary

pigeon breast or the transverse groove round the lower part of the chest from the tug of the diaphragm on the softened ribs of a bronchitic rickety infant, called Harrison's sulcus.

There is a condition met with in both children and adults called resistance to vitamin D in which there is an inability of the body to use normal supplies of the vitamin. The resulting osteomalacia is curable only by the administration of massive doses.

Chronic Steatorrhoea. Steatorrhoea is probably the commonest cause of osteomalacia. Vitamin D is not absorbed because it is fat soluble. Moreover fat retained in the intestine combines with calcium to form insoluble calcium soaps the absorption of calcium as well as that of vitamin D is therefore impaired In the presence of osteomalacia with no obvious dietary abnormality faecal fat analyses are called for Total fat over 25 per cent. of the dried faeces suggests steatorrhoea but this single estimation is of little value. By analysing all stools passed over a 72 hour period the average daily fat excretion can be determined on a normal diet it is less than 6 g. values greater than 10 g. are diagnostic of steatorrhoea. A still more reliable criterion is provided by a 6-day fat balance test loss of more than 10 per cent. of ingested fat is taken as evidence of steatorrhoea. Balance tests, however are possible only in the metabolic ward of a special centre.

There are many possible causes of chronic steatorrhoea, including disturbance of fat digestion from for example chronic pancreatitis, and failure of fat absorption from a number of intestinal diseases. There is also a group whose cause is more obscure coeliac disease in infants and children, idiopathic steatorrhoea, and tropical sprue.

In coeliac disease increasingly frequent attacks of anorexia occur with stools that are bulky and offensive from their high fat content. Loss of weight is progressive and severe. Some of these infants survive with stunted growth and the idiopathic steatorrhoea of adults. Light on the aetiology of coeliac disease came in 1950 with the discovery that the great majority of children recover when the gluten of wheat and rye flour is removed from the diet, and that steatorrhoea promptly recurs when gluten is re introduced.[1]

Idiopathic steatorrhoea and tropical sprue have very similar symptoms, but are probably distinct diseases. There is chronic illness with emaciation, abdominal discomfort with loose offensive stools, recurrent glossitis, sometimes cramp and tetany A macrocytic anaemia develops a normal blood picture is against the diagnosis.

Sprue is a disease to which white races are liable in the tropics. Evidence has grown in recent years that idiopathic steatorrhoea (non-

[1] Ross, C A. C., *et al.* (1955) *Lancet*, i, 1087



FIG. 6a
Osteomalacia pseudo-fractures in the left ulna.

tropical sprue) is adult coeliac disease. The majority of patients are cured by a gluten-free diet, which has no effect on tropical sprue [1]

Symptoms of Osteomalacia. In mild forms the complaint is of pains in the bones of the legs, low backache, and general weakness patients are likely to be sent for examination because of rheumatic pains. In more severe cases, pain, and especially low back pain, may be severe and there is marked general bone tenderness. Muscular weakness is prominent. Skeletal deformities develop. A painful waddling gait from spasm of the adductors of the thigh is fairly common.

The presenting symptom may be an episode of severe back pain which

[1] Cooke, W T (1958) *Brit med. J.*, ii, 261

on X ray examination is found to be associated with a crush fracture of a vertebral body

At times the presenting symptoms are those of tetany [p. 49]

X-ray Appearances in Osteomalacia. Radiographs show a general rarefaction of all the bones of the skeleton with thinning of the cortex, and often pseudo-fractures [FIGS. 6a and 6b] In severe cases gross deformities are common especially in the bones of the pelvis.

Fractures in osteomalacia unite by uncalcified osteoid. Narrow bands

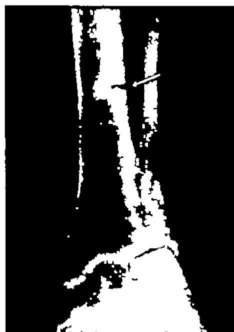


FIG 6b

Another case of osteomalacia showing a well-marked pseudo-fracture on the lower part of the tibia

(by kind permission of Dr F Campbell Golding).

of translucency in the cortex of a bone, transverse to its long axis, are commonly seen in osteomalacia, and in no other condition. They may or may not completely cross the bone and they are usually called pseudo-fractures. They are often symmetrical they may be found in any long bone, but are most common in the femoral neck ischial and pubic rami, the axillary edge of the scapula, and the ribs. The origin of these zones is unknown. Perhaps pseudo-fracture is not appropriate It may be that there is a partial fracture, and the matrix laid down in the crack is not calcified

Milkman's Syndrome Pseudo-fractures may be found when the rest

of the bone appears radiologically normal Milkman s syndrome is this early stage of osteomalacia a case presenting the symptoms of osteomalacia, the serum calcium phosphate and alkaline phosphatase values of osteomalacia, and radiologically pseudo-fractures in a skeleton that appears normally calcified

In this connexion it has been pointed out that before radiologists can detect porosis, some 30 per cent of the bone calcium must have been lost.

Osteoporosis

In osteoporosis inadequate bone deposition is due to a failure of osteoblasts no new osteoid is formed and there is nothing to be re-calcified.

Normal osteoblastic activity probably depends on several conditions, among which are normal hormonal steroids and normal vitamin C. It fails in scurvy in Cushing s syndrome where there is excess of adrenal cortical steroids and at and after the menopause when there is deficiency of oestrogen It fails also in disuse in malnutrition, and in old age. It is a congenital defect in osteogenesis imperfecta. In some instances the reason for osteoblastic failure is quite obscure

There are therefore several clinical types of osteoporosis. In all of them serum calcium, phosphate, and alkaline phosphatase are normal for this is not a disturbance of calcium metabolism.

Menopausal Osteoporosis. Menopausal osteoporosis merging into senile osteoporosis, is a common disorder [FIG 7] It is regarded as generalized, though it affects the spine and pelvic bones mainly long bones much less frequently and skull almost never It occurs in women after the menopause and in men after middle age. It is a common cause of low back pain [p 99]

One of the commoner problems arising in practice is the recognition of a diffuse rarefaction in the spinal column as the common menopausal osteoporosis, and the exclusion of the comparatively rare myelomatosis. There is a stage in the evolution of myelomatosis when X ray films of the spine show no more than this general porosis, and collapse of one or more vertebral bodies. Myelomatosis is also a disorder of late middle age, and the prominent symptom is also pain in the back Although serum calcium is increased in about 50 per cent. of cases, serum phosphate and alkaline phosphatase values are normal. Distinction can be very difficult. Diagnostic criteria are given on p. 97

Scurvy In this condition, there is in addition to the haemorrhagic tendency an osteoblastic failure. The X-ray appearances of the bones in infantile scurvy are characteristic 1 the bones are osteoporotic

cancellous trabeculation cannot be detected, and the shadow of the cortex is reduced to a thin line 2. at the extremity of the diaphysis there is a narrow band of dense bone projecting a little beyond the width of the shaft and this is lying on a narrow completely decalcified zone—a zone responsible for displacements at the growing end, 3 the periphery



FIG. 7
Senile osteoporosis dorsal spine showing compression of vertebral bodies.

of the epiphysis is a sharp thin line, and a faint structureless shadow replaces normal cancellous tissue.

Scurvy is now very uncommon but minor degrees of vitamin C deficiency may contribute to the osteoporosis of malnutrition.

Cushing's Syndrome Osteoporosis is a common finding in Cushing's

syndrome This is a condition due to over-activity of the adrenal cortex and especially to over production of glucocorticoids

Cortisone Therapy When treatment is prolonged especially when the drug is given in doses of the order of 100 mg. daily it has commonly been found to lead to osteoporosis. Collapse of vertebral bodies have often been reported as also have pathological fractures of the neck of the femur the humerus, radius, ribs, and tibia This is now a well known hazard in the prolonged cortisone treatment of rheumatoid arthritis.

Osteogenesis Imperfecta. Osteoporosis from osteoblastic failure occurs as a hereditary defect in osteogenesis imperfecta (fragilitas



FIG. 8
Osteogenesis imperfecta: fracture of the humerus
(by kind permission of Dr Cecil Bull).

ossium). This is a rare hereditary and congenital fragility of bone. It is usually associated with blue sclerotics, and often otosclerosis. All the offspring may be affected or some may have white sclerotics and normal bones. Some of them are stillborn with multiple fractures as a rule fracture from trivial injury begins to occur at about the age of 4 or 5 years—sometimes not until adolescence.

When an ill-nourished child with china blue sclerotics fractures a bone during ordinary activities, the diagnosis is unmistakable. There is none of the tendency found in rickets, to bending of the long bones. X ray films show an egg-shell thinness of the bone cortex, and severe rarefaction

with but few bone trabeculae but never the epiphyseal changes of rickets.

The fractured bones are rarely displaced and unite firmly after adequate fixation [FIG 8] After adolescence fractures may cease to occur but the prognosis is not good from the frequency of fatal inter current disease in the second decade. Many of these children die before adolescence but there is a much less severe form which improves when adult life is reached and which may cause difficulty in diagnosis.



FIG 9
Hyperparathyroidism
(by kind permission of Dr F Campbell Golding).

Hyperparathyroid osteitis fibrosa generalisata

In the general skeletal rarefaction of hyperparathyroidism, bone deposition is normal but it cannot keep pace with the excessive resorption. Hyperparathyroidism is usually due to an adenoma of one of the four parathyroid glands but sometimes to a diffuse hyperplasia of all of them. It affects women more often than men. Most reported cases have been between the ages of 40-50 but instances occur in all decades from the 2nd to the 8th.

The usual symptoms are pain and tenderness in the bones, spontaneous fractures and deformities, and a waddling gait. Pain in back and

limbs may be severe, and the first diagnosis is often rheumatism Besides symptoms referable to the skeleton others are due to disease of the urinary tract, and to hypercalcaemia.

Calcification is likely to occur in the kidneys causing serious changes calcium phosphate calculi with secondary pyelonephritis, or a diffuse nephrocalcinosis leading to renal failure. Hypercalciuria and hyper phosphaturia sometimes cause a marked polyuria and polydipsia, simulating diabetes insipidus Hypercalcaemia causes a decreased muscle excitability and so marked muscular weakness and hypotonicity

X ray films [FIG 9] show a general rarefaction of the whole skeleton and cystic areas of varying size throughout the bone. These cystic areas in the film represent 1 true degenerative cysts 2. areas where bone has been replaced by a vascular connective tissue 3 benign giant cell tumours. Fractures and deformities occur when bones are severely affected deformities of the pelvis, 'fish vertebrae crushed vertebral bodies, and so on. These changes are striking, but not diagnostic almost identical changes may be found in several other bone disorders (v *infra*)

Diagnosis is confirmed by finding a high serum calcium and low serum phosphate, and increased alkaline phosphatase. These bio-chemical changes are almost conclusive.

Differential Diagnosis. The bone changes of hyperparathyroidism are distinguished from osteomalacia and osteoporosis by the X ray appearances and biochemical changes

But very similar X ray appearances are associated with certain localized bone disease, namely polyostotic fibrous dysplasia, multiple myeloma and metastatic deposits. This is the radiologist's problem, and he has two resources 1 he can demonstrate some bone in the skeleton which is normally calcified this excludes hyperparathyroidism 2. he can X-ray the skull—a film of the skull may be pathognomonic in myelomatosis the xanthomatoses and Paget's disease.

The most difficult diagnostic problem—at times insoluble—is to distinguish primary hyperparathyroidism with secondary renal disease from primary chronic renal disease with secondary hyperparathyroidism

Polyostotic fibrous dysplasia (osteitis fibrosa disseminata) is a bone dystrophy of unknown causation, in which the lesions closely resemble those of hyperparathyroidism. But it is not generalized and the serum calcium and phosphate levels are normal.

Myelomatosis [p 94], when multiple osteolytic lesions are present, may closely resemble osteitis fibrosa disseminata radiologically But the lesions are punched out they are much more sharply defined than are the cystic areas of the hyperparathyroid disease. This shows

particularly well in X ray films of the skull. Estimations of serum calcium and phosphate are not of much help for calcium can be high in myeloma, and phosphate, though usually normal can be low Serum alkaline phosphatase, however is nearly always normal. Other diagnostic criteria for myeloma are given on p. 97

The X ray appearances of *metastatic deposits of new growth,* when osteolytic, may be superficially similar to those of hyperparathyroidism but the deposits are much more sharply demarcated Moreover the condition is local or disseminated and not generalized some normal bone will be discoverable Serum calcium may be high, and alkaline phosphatase increased but serum phosphate is very rarely low

Skeletal xanthomatosis

The xanthomatoses, or lipoidoses, are rare diseases of early life, probably congenital errors of metabolism, in which there is excess storage of a lipid in large phagocytic cells— foam cells —in spleen, liver lymph glands and skeleton.

The Hand–Schüller–Christian syndrome is characterized by deposition of cholesterol in typical foam cells, which infiltrate chiefly the bones of the skull, the vertebrae and pelvis, and some of the long bones. The X ray appearances are similar to those of hyperparathyroidism but the condition is localized the serum calcium, phosphate, and phosphatase values are normal.

Renal rickets and renal osteitis fibrosa generalisata

A severe and prolonged renal insufficiency may cause a bone disease in adults indistinguishable from the osteitis fibrosa generalisata of hyperparathyroidism. It is called renal osteitis fibrosa generalisata. In children the same changes may occur with epiphyseal changes in addition a condition called renal rickets, or renal dwarfism

In adults the primary renal disease is chronic nephritis, or chronic pyelonephritis, or congenital cystic disease of the kidney The sequence of events is probably prolonged renal insufficiency → phosphorus retention → low serum calcium → secondary hyperplasia of the parathyroid glands.

But renal failure may supervene in primary hyperparathyroidism from nephrocalcinosis, or pyelonephritis secondary to calculi. Then the previously low serum phosphate is increased, and the previously high serum calcium is decreased. So primary hyperparathyroidism, e.g. from parathyroid adenoma, may produce a picture identical with that of renal osteitis fibrosa generalisata. The only difference is that in the one bone changes came first, in the other prolonged renal insufficiency

COLOUR CHANGES

COLOUR CHANGES IN THE FOOT

Blanching

Raynaud's disease, and the Raynaud phenomenon [p 46] may affect the toes as well as the fingers, although this is far less common.

Severe, continuous, diffuse, and cramp-like pain in the foot and leg is the commonest initial symptom of sudden arterial occlusion It is accompanied and sometimes preceded by numbness and a sense of coldness in the limb On examination, the skin of the foot, and perhaps of the lower part of the leg, is at first blanched with a wax like pallor later it acquires a cyanotic mottling. Prompt diagnosis is vital when a leg suddenly becomes cold and numb and is stricken with severe pain, examine the pulsation in the popliteal and femoral arteries.

Cyanosis

A cyanotic tint in the skin indicates as a rule a sluggish blood flow through widened capillaries, due to partial arterial obstruction or to venous obstruction Thus cold blue feet are found in

Arterial Occlusion. Occasionally the feet, as well as the hands, are involved in acrocyanosis, a condition due to over-constriction of arterioles in cold weather Varying degrees of arterial occlusion occur in thrombo-angiitis obliterans and in arteriosclerosis obliterans. The first sign of arterial disease may be discoloration of the foot and leg—commonly a cyanotic tint of the toes and dorsum of the foot. A cold pale cyanotic skin indicates a very sluggish or absent blood flow

Sometimes the skin of the foot is cold and red This may be so when the leg is dependent or when the temperature of the foot is so low that oxyhaemoglobin will not part with its oxygen.

Venous Occlusion. Iliofemoral thrombophlebitis results in rapid and marked enlargement of the whole lower limb with distension of the superficial veins and cyanosis of the skin The residual chronic venous insufficiency is characterized by oedema of the leg, the skin of which is usually but not always cyanosed When venous insufficiency is of long standing, brownish pigmentation appears in irregular areas in the lower part of the leg.

Disuse Use of a limb increases the resting blood flow through the tissues disuse has the reverse effect. If movements of the foot are restricted, say by joint disease, its temperature falls, and the continually cold foot becomes more and more cyanotic.

Disuse may be the result of any chronic painful condition of diseased joints, or of paralysis from disease of the central nervous system. The cyanosis of disuse is particularly well seen in the residual paralysis of poliomyelitis foot and leg are continually cold, the skin is mottled with red purple, or cyanotic patches, and chilblains are common. Atrophic changes in the skin—and in all the structures of the disused limb—gradually follow on the defective circulation and the limb may be slightly oedematous.

For the same reason, a cold blue foot and leg is a consequence of spastic paralysis of the lower limb

Cold red feet and legs

Chronic Pernio (Erythrocyanosis). With repeated exposure of feet and legs to cold, in the winter months, chronic chilblains develop appearing in successive crops, and tending to ulcerate. Ulcerated lesions heal in 3-5 weeks, leaving a pigmented scar The condition occurs almost exclusively in women beginning in adolescence or early adult life. The lesions appear on the lower third of the leg, on anterior and posterior surfaces, and often on the dorsum of the foot. They may be confluent, forming a single irregular area of reddened or cyanotic tender skin, hard and raised, and with here and there ulcerating nodules.

Chronic pernio is either confined to the winter months, or if it does not disappear in the summer it improves considerably This is an important diagnostic point but not quite pathognomonic.

Erythema Induratum (Bazin's Disease) This is the condition most likely to be confused with chronic pernio Cases of chronic pernio as well as of erythema induratum have been classified as Bazin's disease but erythema induratum occurs independently of the seasons. Subcutaneous nodules are found on the legs, usually the lower part of the calves, covered with a bluish red or ulcerated skin. The nodules vary from about 1 to 3 cm., but are often confluent. They are usually painful and tender and are very chronic, persisting for years There is a good deal of induration around ulcerated nodules.

They are commoner in women than in men the age incidence is 10-40 years. Some instances are tuberculous in origin, associated with tuberculous adenitis

Livedo Reticularis. The skin of the feet and legs (and hands and arms) is marked by reddish blue reticulation, or mottling. Discoloration is more pronounced in cold, less in warm weather It affects young adults, who may have no symptoms, but who may complain of coldness, numbness, and a dull aching in legs and feet. The condition is due to constriction of certain of the skin arterioles

arterial pulsation is normal It is commonly associated with chronic pernio

Painful red feet

Erythralgia This names a reddened skin which is extremely painful when stimulated by friction or moderate warmth—i.e. a temperature conveying to normal skin a sensation merely of warmth A skin inflamed from any cause has these properties.

Erythromelalgia (Erythermalgia). This name is given to a clinical syndrome characterized by erythralgia in chronic form affecting the feet (p. 114)

COLOUR CHANGES IN THE HAND

Blanching

A blanched skin, or a pale violaceous tint, indicates complete, or almost complete, arrest of blood flow Many people are liable to attacks of dead fingers in cold weather due to temporary cessation of blood flow through the digital arteries.

In mild attacks one or two fingers only are affected but vasospasm may be extensive involving most of the fingers in both hands, sometimes the whole hand—even hand and lower forearm

The sufferer may have continually cold, slightly cyanotic hands, liable to chilblains. In an attack the skin of a finger becomes pale and waxy first at the tip, spreading up the finger to a variable extent. If vasospasm is prolonged the finger becomes numb and painful. The cause of this attack—which is called the Raynaud phenomenon—is spasm of the digital arteries, and of the minute skin vessels. Recovery is marked first by relaxation of the minute vessels, into which blood flows from the veins giving the skin a cyanotic tint then, with relaxation of the digital arteries, rubor appears at the base of the finger and spreads to the tip

The Raynaud phenomenon does not occur in normal people cold causes contraction of digital arteries, but not sufficient to arrest the blood flow It may occur as a symptom, when the lumen of the arteries is narrowed by disease or it may occur in the absence of any discoverable primary condition, in which case alone should we call it Raynaud's disease.

Raynaud's Disease This is a fairly common familial disorder in girls and young women. Attacks of the Raynaud phenomenon are nearly always bilateral. They occur in response to exposure to cold, but are sometimes precipitated by emotional excitement. Usually the attacks

are mild and in the intervals the fingers either present no abnormality or are continually cold and subject to chilblains.

In some cases, however attacks are frequent, extensive, and prolonged In these sclerodactyly [p. 194] may develop The skin becomes smooth and fixed to underlying tissues, and flexion of the fingers is restricted.

Raynaud's Phenomenon. This is a safer label at least for a year or two until one can be quite sure that there is no evidence of thrombo-angiitis, or other cause. It may be secondary to

1 occlusive arterial disease, especially thrombo-angiitis obliterans
2 the cervical rib or scalenus, syndrome
3 the use of pneumatic hammers—principally by stone cutters and riveters.

Thrombo-angiitis Obliterans [p 145]. This disease is suspected when unilateral attacks of Raynaud s phenomenon occur in a young man. In this disease the arteries of the upper extremities are involved in about 40 per cent. of cases and because attacks of blanching of the fingers may be a prominent feature in the early stages, the arterial disease is often confused with Raynaud s disease. Both occur in young people, but the diagnostic points shown in the table are helpful.

	THROMBO-ANGIITIS	RAYNAUD'S DISEASE
Sex	males 98 per cent.	females 80 per cent.
Symmetry	at first unilateral later bilateral but asymmetrical	bilateral and symmetrical
Recurrent phlebitis	common	none
Arterial pulsation	may be absent	normal
Postural colour changes	often present	none

The Cervical Rib (or Scalenus) Syndrome [p. 81] Although in most instances pain in the arm is the prominent symptom, and symptoms due to pressure on the axillary artery are trivial, prolonged attacks of Raynaud s phenomenon may occur the hand tending to be cyanotic in the intervals. Permanent coldness of one or more fingers is suggestive of arterial occlusion, and the cervical rib syndrome in young men is therefore liable to confusion with thrombo-angiitis obliterans. But the cervical rib syndrome is more common in young women.

Pneumatic Hammers When Raynaud s phenomenon occurs in a man, it is well to inquire about the use of percussion tools. For this is a well known occupational disease repeated percussion so affecting the digital arteries that they become hypersensitive to cold. Before assuming

that the condition is occupational, however, thrombo-angiitis obliterans and the cervical rib syndrome must be excluded

Blanching of the hand associated with numbness and severe pain occurs in sudden arterial occlusion this is much more common in the lower limb [p. 43]

Cyanosis

A cyanotic skin usually indicates a sluggish blood flow through dilated capillaries. This assumes that the skin is supplied with normal arterial blood.

Cyanosis of both hands is seen in the very common condition called acrocyanosis. This is the result of hypersusceptibility to cold, of arterioles their over-constriction slows the blood flow in the capillaries which become very dilated. The cold skin is cyanotic, or red if very cold or a blotchy bluish purple. Cyanosed hands become bright red when they are so cold that oxyhaemoglobin will not part with its oxygen Acrocyanosis is closely allied to Raynaud s disease, in which a similar fault affects the digital arteries.

The condition is found mainly in young women. Their hands in the winter months are cold and cyanosed The colour changes to bright red when the hands are very cold—or when they are warmed Both hands are equally affected, and occasionally the feet also chilblains are common. Arterial pulsation in the limbs is normal there is no arterial disease.

Both hands may be cyanosed in patients who have suffered for many years from Raynaud s disease.

A cold cyanosed hand in one limb only is usually a consequence of disuse, from pain weakness, or joint stiffness. Disuse results in a fall of temperature sufficient to give a cyanotic tint to the skin. The chief causes are 1 polyarthritis 2. atrophic paralysis of the hand, e.g. progressive muscular atrophy syringomyelia, etc. [p. 17] hemiplegia.

In the cervical rib syndrome the hand is not infrequently cold and cyanosed, and perhaps swollen. This may be due simply to disuse of a weak painful limb or in part to pressure on the subclavian artery

Cyanosis of one hand may also be caused by venous obstruction, as in the uncommon axillary and subclavian thrombophlebitis.

The hand is noticeably cold and cyanosed in the reflex sympathetic dystrophies [p 83]

CRAMP

Cramp is an involuntary sustained muscular contraction, usually severely painful. Most of us have had episodes of cramp after severe exertion. But we are concerned here with cramp coming on during ordinary activities, or when resting. It is due to increased irritability of the neuromuscular mechanism the cause of which is sometimes known, more often quite obscure.

UNKNOWN AETIOLOGY
Nocturnal cramps in the elderly
Professional cramps
KNOWN AETIOLOGY
Salt deficiency
Tetany
Nervous diseases

Nocturnal cramps

These, especially in the elderly are fairly common. Very occasionally one of the conditions discussed below will be found responsible usually there is no demonstrable cause. In most instances it is not a defective circulation, for subjects of arteriosclerosis rarely complain of nocturnal cramp.

Professional cramps

Such cramps are apt to occur in persons doing any form of repetitive manual work—writers, typists, pianists, violinists, and so on. They are becoming uncommon.

Writer's Cramp This is typical of the group After writing for a variable time—sometimes only a minute or so—movements become jerky then cease from painful cramp of the hand and forearm muscles. The distinguishing feature is that only writing produces the cramp never any other use of the hand. One should be careful, however to exclude local disorders of the hand and wrist, and early organic nervous disease—especially Parkinson's disease. There is no good reason for calling the condition a neurosis it probably arises from faulty co-ordination of the hand and forearm muscles used in writing.

Cramp and salt deficiency

Sweat is hypotonic saline. If copious sweating is immediately followed by drinking hypotonic saline, a normal salt concentration in the interstitial fluid is restored. But if thirst is relieved by copious draughts of water though the volume of interstitial fluid is made good, it becomes hypotonic. Osmotic equilibrium between extra and intra-cellular fluid

is then restored by a shift of extra-cellular water into the cells. The cells become hydrated This appears to be one cause for increased irritability of muscle cells, and for the condition that brings on severely painful cramps, called miners cramp stokers cramp or heat cramp.

Under normal conditions this disturbance in distribution of body water is soon corrected but this is not possible in conditions of marked salt deficiency It has been shown experimentally that a salt free diet brings a tendency to cramp In practice, no evidence of salt deficiency will be found in most sufferers from cramp, nor any relief obtained by administration of salt. But a few cases have been reported of cramp undoubtedly so caused—e.g. after many years on a salt free diet—and the possibility is worth remembering.[1]

Tetany

Tetany is a syndrome due to increased irritability of motor and sensory nerves, both somatic and vegetative resulting from 1 a decrease in the ionized calcium of the serum or 2. a decrease in the H-ion concentration of the serum There is recent evidence that the immediate cause may be the increase in the serum phosphorus that accompanies hypocalcaemia Probenecid relieves the tetany of hypoparathyroidism as soon as the serum phosphorus level is reduced to normal, and before there is any perceptible rise in the serum calcium level.[2]

The prominent symptom is painful paroxysmal cramp of the extremities, or in severe cases of almost the whole skeletal musculature. Cramp may be limited to hands and feet—carpopedal spasm fingers and thumbs drawn together into a cone (*main d'accoucheur*) feet held stiffly in equino-varus. But it may spread to the rest of the limbs wrists and elbows flexed and arms folded, legs extended. In very severe cases spasm may become generalized, the body being rigidly extended for minutes or hours.

Increased irritability affects also the vegetative nervous system spasm of the intestinal tract causing pain and vomiting of the bladder retention or incontinence of peripheral blood vessels, pallor of the digits of the heart, sudden death. Increased irritability of sensory nerves is manifest by a complaint of tingling or burning paraesthesia in the extremities, associated with the painful cramps.

Spontaneous cramps follow a variable period of latent tetany during which increased neuromuscular excitability can be detected by the following tests

1 *Chvostek's sign.* A light tap over the facial nerve, just anterior to

[1] Hall, A (1947) *Lancet*, ii, 321
[2] Beidleman, B. (1958) *Metabolism*, 7 690.

the external auditory meatus, causes a twitch of the facial muscles on that side.

2. *Trousseau's sign.* If the circulation in one arm is stopped for about 3 minutes, by inflating a blood-pressure cuff a typical carpal spasm results.

The causes of tetany are the causes of hypocalcaemia, and the causes of alkalosis. Hypocalcaemia sufficient to cause symptoms of tetany may occur in the following disorders

Hypoparathyroidism. Most cases are post-operative, the result of accidental removal of parathyroids, or injury to their blood supply during thyroidectomy There is also an idiopathic type which is very rare. Tetany may occur when the serum calcium falls to 4-8 mg. per 100 ml.

Infantile tetany is usually associated with rickets. The serum calcium in rickets, and in the osteomalacia of adults, is usually normal. But in some cases, especially if calcium intake is deficient, it is low and symptoms of tetany may appear

In pregnancy and lactation the serum calcium occasionally falls sufficiently to cause mild cramps, and tingling, burning paraesthesia.

Alkalosis There is no clinical difference between the tetany of hypocalcaemia and that of alkalosis. A decrease in the H ion concentration causes increased neuromuscular irritability as does hypocalcaemia. Probably the immediate cause is the same a reduction in the ionized calcium of the serum, although the total calcium is normal. Alkalosis occurs in

1 *Hyperventilation* overbreathing—usually a neurotic manifestation.

2. *Prolonged vomiting* as in pyloric obstruction It is due to excessive loss of hydrochloric acid.

3 *Excessive alkali intake* a possibility in the alkali treatment of peptic ulcer

Tetany must be distinguished from tetanus which occurs from 1 to 6 weeks after infection of a wound by the tetanus bacillus. The wound, no more than a small puncture, may have been overlooked. The course of the disease is characteristic stiffness of the neck and lock jaw (trismus), followed within a few days by a general muscular rigidity and paroxysms of violent spasms in all skeletal muscles, lasting a few seconds and recurring at varying intervals. They may be distinguished from the spasms of strychnine poisoning by the absence of complete relaxation in the intervals. The trismus of tetanus is soon distinguished from that due to an impacted wisdom tooth, or to hysteria, by the appearance of general rigidity

Cramp in nervous diseases

Cramp may be a comparatively unimportant symptom in a number of diseases cramp in the toes in paralysis agitans, for instance or recurrent cramps in the calf and plantar muscles in sciatica. Bilateral calf and plantar cramp especially with foot drop or paraesthesia, suggests multiple peripheral neuritis.

DEFORMITIES

DEFORMITIES OF THE FOOT

OF THE TOES
- *Hallux valgus*
- *Claw foot*

OF THE REST OF THE FOOT

CONGENITAL	ACQUIRED
Talipes equinovarus	*foot drop* (*pes equinus*)
Talipes calcaneovalgus	*pes cavus*
	pes valgus

Hallux valgus

The first abnormality in the development of hallux valgus is a widening of the interval between the first and second metatarsals—a varus deformity of the first metatarsal Why this happens is unknown, and probably different factors operate in different cases. The condition is often familial, first appearing in childhood or adolescence The big toe is prevented from following its metatarsal to the new position by the pull of muscles and the restraint of footwear It becomes deflected towards the other toes [FIG. 10]. This valgus deformity gradually increases, sometimes so much that the toe eventually lies transversely across the others It may lie on the dorsum of the other toes, or some times under the second toe which becomes clawed and subluxated

The prominence of the first metatarsal head is increased by the formation of osteophytes, or an exostosis, and by an inflamed bursa over this. Sooner or later osteo-arthritic changes appear in the meta tarso-phalangeal joint. The other metatarsal heads are depressed, the ball of the foot becoming more or less concave and its skin thickened by callosities.

This is the commonest deformity of the toes. It is found in women much more often than in men.

Claw foot

Clawing of the toes arises essentially from the same condition that

causes claw hand namely paralysis of the interosseus and lumbrical muscles. Their action is to extend the interphalangeal joints and flex the metatarso-phalangeal joints Unopposed action of the long flexors on the interphalangeal joints and of the long extensors on the metatarso-phalangeal joints will therefore produce a claw like deformity of the toes. This deformity is commonly associated with pes cavus. The not

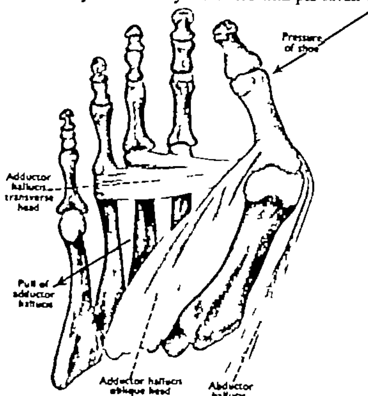


FIG. 10
Hallux valgus. The head of the first metatarsal has become widely separated from that of the second In the valgus position of the phalanges the pull of the adductor exceeds that of the abductor hallucis.

uncommon idiopathic pes cavus and claw foot begins in childhood. Possible though rare causes in children are peroneal muscular atrophy [p. 9], and Friedreich s disease [p 7].

When the deformity first appears in adult life, the cause may be

1 Reflex atrophy of the intrinsics from rheumatoid arthritis affecting the foot.

2. Paralysis of the intrinsics from a nerve lesion. This is very uncommon, and the possibilities are fewer than for the corresponding condition in the hand. Injury of the tibial, or external plantar nerve is rare so also is a root lesion of S.2. It is a possible residual paralysis of acute poliomyelitis.

Talipes equinovarus

The commonest congenital club foot results from a combination of plantar flexion and inversion, occurring at the ankle, the sub-taloid and the mid-tarsal joints. At first with some force passive restoration of a plantigrade position is possible. But as time goes on changes in the shape of the bones, and contractures, bring increasing and persisting deformity

Talipes Calcaneovalgus. This is a deformity compounded of extreme dorsi-flexion at the ankle—the dorsum of the foot in contact with the leg—and eversion at the sub-taloid and mid tarsal joints.

Foot drop (PES EQUINUS OR EQUINOVARUS)

Normally the foot can be dorsiflexed beyond the plane at right angles to the leg. The dorsiflexors are the tibialis anterior extensors of the toes, and peronei supplied by the common peroneal nerve (L.4 L.5 S 1)

Apart from congenital deformity and one due to injury or arthritis, foot drop is the result of upper or lower motor neurone paralysis of these muscles. The foot cannot voluntarily be dorsiflexed to a right angle the toes drag along the ground in walking, repeatedly tripping over slight obstructions.

FOOT DROP FROM UPPER MOTOR NEURONE PARALYSIS

In upper motor neurone paralysis of the lower limbs, the dorsiflexors of the foot are affected earlier and more severely than the plantar flexors and other muscle groups. The reverse holds for spasticity the plantarflexors become more spastic than the dorsiflexors Pes equinus is therefore an early symptom of spastic paralysis of a lower limb from a lesion of the pyramidal system. The causes are reviewed on p. 212.

FOOT DROP FROM LOWER MOTOR NEURONE PARALYSIS

Anterior horn cells
- Acute poliomyelitis [p 8]
- Progressive muscular atrophy (rare) [p 10]
- Peroneal muscular atrophy [p. 9]

Nerve root
- Sciatica from root compression [p 163]

Peripheral nerves
- Neuritis of the common peroneal nerve
- Multiple peripheral neuritis [p 14]
- Compression of sacral plexus by pelvic tumour

Neuritis of the Common Peroneal Nerve The peroneal nerve may be compressed or injured where it winds round the upper end of the fibula

or it may be affected by a neuritis of unknown causation. A severe foot drop follows a few days of pain in the lateral side of the leg. Some wasting develops in the paralysed peronei, tibialis anterior and extensors of the toes and there is a variable sensory loss in the overlying skin. Recovery is slow and usually incomplete.

Closely similar but usually much less severe is the weakness and wasting that may develop in sciatica. The foot drop of multiple peripheral neuritis is readily distinguished by the absence of objective signs of sciatica, by being bilateral and by impaired sensation in stocking areas.

Pes valgus

The Longitudinal Arch of the Foot The medial side of the foot, more massively built than the lateral, is designed to carry most of the body weight the lateral part is more concerned with stability and balance. The bones of the medial part—calcaneus, talus, navicular cuneiforms and inner 3 metatarsals—form an arch from the heel to the ball of the foot.

The arch is prevented from collapsing under the weight of the body not by the shape of the bones, but chiefly by the tibialis posterior the tendon of which passes beneath the calcaneo-navicular ligament to its insertion into the plantar surface of the small tarsal bones. To a less extent the tibialis anterior has a similar action.

The calcaneo-navicular ligament, the ligaments of the individual tarsal joints, and the long and short plantar ligaments of the foot, could not alone prevent the arch from collapsing under the weight of the body And this is not the function of ligaments When these ligaments are subjected to strain, a reflex is initiated which ends in contraction of the invertor muscles and relief of the strain. When the muscles fail and strain comes on the ligaments alone, chronic ligamentous strain and a gradually increasing valgus deformity of the foot result.

Movements of the Foot Apart from ankle and toe movements, these are 1 a very restricted range of dorsi- and plantar flexion 2 the compound movements of inversion and adduction in one direction, eversion and abduction in the other The compound movements occur at the talo-calcanean and talo-navicular joints. Inversion and adduction cannot occur separately and the movement may be referred to simply as inversion nor can eversion and abduction which may be referred to simply as eversion. Inversion of the foot raises the arch eversion lowers it. A rotary movement imparted to the talus with the foot stationary achieved by rotating the lower limb at the hip while standing, has the effect of raising or lowering the arch, in moderate degree external

rotation inverts the foot and so raises the arch Internal rotation has the opposite effect

Pes valgus is a foot that has assumed the everted position, either when off the ground or only when weight bearing. It is easy to recognize when the patient stands the inner border of the foot rests on the ground or almost so, and its normally straight or slightly concave outline has become convex. Clearly a foot may do this for a number of reasons. The following causes of pes valgus are known or have been suggested

1 A general bad posture, or muscular atonia the immediate cause being internal rotation of the lower limbs from weak gluteal muscles.

2. Extension deformity of the first metatarsal the foot to become plantigrade when standing must go into the valgus position.

3 A short tendo Achillis the heel can only make contact with the ground—barefoot or in low heeled shoes—if the foot goes into the valgus position.

4 Weakness of the invertors of the foot—the tibialis muscles—so that the strain of the body weight is taken by the ligaments of the arch.

5 Normal tibialis muscles submitted to an excessive strain—from increasing body weight, or unaccustomed long hours of standing—so that again the strain comes on the ligaments of the arch.

6 Spasm of the evertors of the foot—the peroneal muscles—spastic flat foot.

7 Arthritis of the tarsal joints—more importantly rheumatoid arthritis

8. Osteo-arthritic changes in the tarsal joints secondary to prolonged foot strain.

9 Congenital flat foot.

Two varieties of pes valgus need a special note.

In *spastic flat foot* the foot is rigidly everted by spasm of the peroneal muscles it is most common in adolescent boys, and very rare in adults. An arch can be reformed when the spasm relaxes under anaesthesia. Soon after leaving school and beginning work involving much standing, the boy complains of pain in one or both feet, gradually increasing in severity The cause of the spasm is unknown. Often it is associated with calcaneo-navicular fusion—a congenital bony abnormality

Congenital flat foot is a rare disorder The degree of valgus deformity increases with growth, and in adolescence or early adult life the pain and clinical signs of spastic flat foot appear

The cause is a congenital bony abnormality a bridge of bone joining the sustentaculum tali to the talus, preventing any movement at the

talocalcanean joint, and forcing the talus into an abnormal position, with its head bulging the inner border and sole of the foot.

It is distinguished from other varieties of spastic flat foot by the fact that the foot is rigid from birth, the arch cannot be reformed even under anaesthesia, and the deformity increases throughout childhood.

DEFORMITIES OF THE HAND

Claw hand

Claw hand is the deformity of hyperextension at the metacarpo-phalangeal joints, and flexion at the interphalangeal joints. It results whenever the intrinsic muscles of the hand are paralysed while the extensor digitorum communis is intact.

The interossei are the adductors and abductors of the fingers. Also, with the lumbricals, they flex the first phalanges and extend the second and third phalanges. The extensor digitorum communis is believed to act almost entirely on the first phalanges.

Partial claw hand, in which the ring and little fingers are mainly affected, is found in ulnar paralysis [p 11] and in the ulnar type of cervical rib syndrome [p 81]

The full claw hand develops at some stage in the progressive paralysis of syringomyelia [p 16], and of progressive muscular atrophy and amyotrophic lateral sclerosis [p 10] Atrophy of the intrinsic muscles of the hand from compression of C.8 and T 1 anterior nerve root will also produce this deformity—provided C.7 root supplying the extensor digitorum communis is missed Bilateral claw hand and claw foot in an adolescent is characteristic of peroneal muscular atrophy [p. 9].

Clubbed fingers

Enlargement of the tips of the fingers (and toes) from thickening of the soft tissues of the terminal phalanges, with increased longitudinal and transverse curving of the nails, usually occurs alone, but may be associated with hypertrophic osteo-arthropathy [p 205]. The bone and joint changes of this condition arise from the same causes as clubbing, but are much less common.

Clubbing may exist as a familial peculiarity in perfect health. Usually it is pathological, the main causes being

- Cyanotic congenital heart disease.
- Chronic pulmonary diseases fibrosis of the lung with bronchiectasis bronchial carcinoma.
- Minor degrees may be found in emphysema, mediastinal pressure, and mitral stenosis.

Flexion deformity of the fingers

The causes of inability to achieve full extension of the fingers may be grouped as follows

Paralysis of extensors radial paralysis [p. 11]

Paralysis of intrinsic muscles claw hand [p 56]

Muscle lesions Volkmann s ischaemic contracture

Skin lesions sclerodactyly [p. 194]

Fascial lesions Dupuytren s contracture

Joint lesions rheumatoid arthritis some instances of osteo-arthritis

Tendon and tendon sheath lesions injuries— mallet finger button-hole deformity rheumatoid nodules in the tendons of the palm gout—uric acid deposits in the tendons Congenital flexion of the little finger

Volkmann's Ischaemic Contracture This condition results from occlusion of the brachial artery from injury The mechanism is often reflex arterial spasm. It may follow injury to the elbow in children, most frequently a supracondylar fracture of the humerus. The flexor muscles of the fingers and wrist become so ischaemic that some of the fibres necrose. Necrotic fibres are later replaced by fibrous tissue and the muscles shorten.

Within a few hours of the injury there is severe pain in the hand and fingers. The hand quickly becomes cold, cyanosed and swollen, and the fingers flexed. Swelling abates, but the flexion deformity increases. It can be observed that extension of the fingers is possible if the wrist is flexed, but that extension of both is impossible showing that the flexion deformity is due to shortening of the flexor muscles

Dupuytren s Contracture This affects chiefly men after middle age, and is very easily recognized. One or more, and sometimes all of the fingers are pulled into flexion by a progressive thickening and con tracture of the palmar fascia. Seen early the skin of the palm is puckered and contains one or more hard nodules. The ring finger is flexed at the metacarpo-phalangeal and proximal interphalangeal joints, and attempts to straighten it are clearly resisted by a taut palmar fascia. The other fingers follow and after months or years the tips are pulled hard against the palm.

Tendon and Tendon Sheath Lesions Flexion deformity from tendon injury is not difficult to recognize voluntary extension is lost, but passive extension is normal. Rupture of the extensor pollicis longus tendon over the dorsal surface of the lower end of the radius may follow fracture and may occur spontaneously in rheumatoid arthritis. Voluntary extension at the interphalangeal joint is lost.

Mallet finger results from rupture of the extensor tendon at its insertion into the base of the terminal phalanx voluntary extension at the terminal interphalangeal joint is lost, and the phalanx is held in flexion. In the so-called *button-hole deformity* the central part of the extensor tendon, which is inserted into the middle phalanx is ruptured the finger assumes a position of flexion at the proximal interphalangeal joint, and cannot be extended here.

Nodules and thickenings in the flexor tendons and sheaths of the fingers, causing flexion deformity are very common in rheumatoid arthritis, at times with little or no joint involvement. The tendovaginitis of this disease may be extensive [p. 127].

Flexion deformity may occur from deposits of uric acid in the flexor tendons and tendon sheaths in chronic gout.

Hereditary deformities of the hand

The many hereditary deformities of the hand present no diagnostic problem. The following may be briefly mentioned

Radio-ulnar Synostosis The upper part of the radius and ulna are fused. The hand is held mid way between pronation and supination, and rotary movement is impossible.

Congenital Absence of the Radius This is the commonest of many deformities due to absence of a bone. The hand is deviated laterally and may be at right angles to the ulna.

Flexion Deformity of the Little Finger This condition and also lateral deviation at one of the interphalangeal joints of the little finger or index are very common hereditary defects.

Syndactylism and Webbed Fingers. Adjacent fingers are joined by a web of skin extending from the base any distance to the tips.

DEFORMITIES OF THE KNEE

Genu valgum (KNOCK KNEE)

Idiopathic In the first 5 years of childhood some degree of knock knee is very common. The medial femoral condyles have grown more rapidly than the lateral. This unequal growth is corrected later and in the great majority of these children the legs are straight again by 6 years.

The degree of genu valgum in children is measured by the separation of the medial malleoli when the knees are together If this amounts to as much as 3 inches at the age of 3 years, spontaneous correction is doubtful and continued increase above 3 inches after this age is taken as an indication for operation.

Due to Bone Disease The commonest disease of bone to cause knock knee in children is rickets. In children and in adults it may appear as a

result of other rarefying diseases of bone [p 31] In adults the deformity may be the result of severe arthritis of the knee—either osteo-arthritic, or rheumatoid in type.

Genu varum (BOW LEGS)

The usual cause of an exaggerated curve of the legs in former times was rickets. In children, the cause may be unequal growth of the femoral condyles, a defect similar to that responsible for idiopathic knock knee, and one which is soon naturally rectified. Appearing in adults, apart from the generalized rarefying diseases of bone, the cause will be found in deformity of the femoral or tibial condyles from chronic arthritis of the knee joint.

INVOLUNTARY MOVEMENTS

There should be no difficulty in placing an involuntary movement in one of the following categories

CONFINED TO A SINGLE MUSCLE OR PART OF A MUSCLE
Fibrillation
Myoclonus

RHYTHMIC ALTERNATE CONTRACTIONS OF MUSCLE GROUPS AND THEIR ANTAGONISTS
Tremor

IRREGULAR MOVEMENTS
Chorea
Athetosis

REPEATED CO-ORDINATED MOVEMENTS
Tics habit spasms spasmodic torticollis

FIBRILLATION

This is an involuntary quivering of a part of a single muscle. It may occur in fatigue, and in ill-health from any cause. It is a sign of diagnostic value in that it occurs in muscles that are wasting from a slow degeneration of anterior horn cells, especially in progressive muscular atrophy [p. 10] It is either a spontaneous quivering, or can be elicited by gently tapping the muscle.

MYOCLONUS

Myoclonic movements are sharp brief spasms, mild or severe affecting one or more muscles, often symmetrically on the two sides of

the body It is a rare disorder apart from its association with epilepsy In myoclonus epilepsy epileptics are affected by myoclonic contractions in the intervals between epileptic attacks. In paramyoclonus multiplex the involuntary movements appear in adult life, with no clue to their causation. Weak contractions regularly repeated affect muscles of upper and lower limbs.

TREMOR

Rhythmic contractions of a muscle group and its antagonist, alternately are called tremor The involuntary oscillation has a frequency of 3 to 9 per second Its amplitude divides tremors into fine and coarse—a distinction of little diagnostic value. A simple tremor affects one muscle group and antagonist only a compound tremor affects several groups, and the movement is complex Intention tremor is described below

Tremor is not always a sign of disease. It occurs in fear and other strong emotions and in muscle fatigue from any cause Also of no significance is familial tremor a fine tremor of the hands in children and young adults, tending to disappear in later life. There are no symptoms and signs of disease, and a family history will probably be obtained.

A fine and rapid tremor of the fingers, and sometimes of the facial muscles, is a common and early sign of chronic alcoholism the recognition of which is always important. Other signs and symptoms are a general restlessness, morning vomiting, diarrhoea, the well-known coarseness of the face with its telangiectases, and loss of memory for recent events.

Diseases in which tremor is often prominent are thyrotoxicosis, Parkinsonism and disseminated sclerosis and in considering their differential diagnosis we have to remember that any type of tremor may be seen in the psychoneuroses.

Thyrotoxicosis

The tremor is a fine rapid oscillation of the fingers when the hands are outstretched. It differs in no important way from the tremor of hysteria, of anxiety or of alcoholism. Thyrotoxicosis affects mainly females, from puberty to middle age. The classical signs are enlargement of the thyroid, exophthalmos and other eye signs, tremor and tachycardia. But the diagnosis may rest merely on tremor and tachycardia (a pulse rate of over 120) in a patient giving a history of gradually increasing fatigue, shortness of breath, loss of weight, irritability excessive sweating, intolerance of heat, and indifference to cold.

In doubtful cases the diagnosis may be confirmed by an estimation of the basal metabolic rate (B M R.) This is usually increased to between

+30 and +75 per cent above the normal standard. A more reliable test is to measure the B M R. before and after a 2 weeks course of iodine A significant fall is characteristic, and is not found with any other cause of an increased B M R.

Very strict attention to detail is needed in the preparation of a patient for a B M R. test unless mental and physical calm are assured the result is of no value. Up to 15 per cent. above, and down to 15 per cent. below normal is regarded as the normal range.

A more reliable indication of thyroid activity—except in either surgically or medically treated cases, when the B M R. is superior—is given by an estimation of the rate of radioactive iodine uptake by the thyroid gland after oral administration. The patient is given a dose of iodine 131 in aqueous solution by mouth and returns 3-4 hours and again 24 hours later for determinations of its uptake by the gland. This test can be carried out only at special centres.

Parkinsonism

The tremor is of two kinds 1 fine oscillations in the extremities and head 2. coarse slow compound movements— cigarette rolling movements of the thumb and index alternate flexion and extension at the wrist, or pronation and supination of the forearms. It is most marked when the limb is at rest, inadequately supported When severe it may continue during sleep. It is increased by emotion and lessened by voluntary movement.

Parkinsonism is a syndrome caused by degenerative changes in the extrapyramidal motor system, most cases being either idiopathic paralysis agitans (Parkinson s disease) or post-encephalitic—a sequel of encephalitis lethargica.

Paralysis agitans affects males more often than females, first appearing in the 6th or 7th decade. The salient features are tremor and muscular rigidity one or the other of which may greatly predominate. In an advanced case the disease is obvious at a glance a patient with a mask like face, stooping posture, tremulous arms held slightly abducted at the shoulder and flexed at the elbow shuffling along with short steps, tending to accelerate and break into a little trot. Many years will have elapsed before this stage is reached and at the onset diagnosis may be quite difficult.

The earliest complaint is often a loss of dexterity in one hand [see writer s cramp p 48] The hand may show a tremor By repeatedly flexing and extending the wrist the examiner may detect rigidity in the muscles of the forearm, of the type called lead pipe or cog-wheel Tremor and rigidity are at first unilateral, remaining in the arm and leg

of one side perhaps for some years. One of the earliest signs is that one arm has lost its natural swing in walking. The face becomes more and more expressionless, and normal fleeting changes of emotion are not registered It happens now and then that an early case of paralysis agitans is considered to be suffering from rheumatism for complaint is often made of aching pain in the shoulders and arms, of a heaviness in the legs, and of cramps in the feet.

Other causes of tremor

Senile tremor is a fine tremor of hands and arms and head, but is always bilateral it is not associated with muscular rigidity nor any of the other features of Parkinsonism

Tremor occurs in several forms of gross cerebral disease some cerebral tumours occasionally in the paralysed limbs of hemiplegia, in progressive lenticular degeneration—a disease of the old motor system a feature of which is the Parkinsonian type of tremor and in dementia paralytica.

Intention Tremor In the main this affects the upper limbs, appearing during the performance of some voluntary action. It can be best elicited by holding out one of the patient s arms and asking him to bring the index finger to the tip of his nose or by asking him to touch an object placed at arm s length from him. The intended movement is marred by oscillations of the limb regular or irregular, increasing in amplitude as the goal is approached the index may rest oscillating for a time on the tip of the nose. This type of tremor usually indicates interference with cerebellar postural control but it may also occur in hysteria. Of the organic diseases of which intention tremor is a feature, disseminated sclerosis is by far the most common [p. 212] Others are Friedreich s disease and a number of rare cerebellar lesions.

CHOREIFORM MOVEMENTS

Choreiform movements occur in rheumatic chorea (Sydenham s chorea) Huntington s chorea, and hysteria.

Rheumatic chorea

This affects children, from 5 to 15 years, girls more often than boys. Rare instances are encountered in adult life, usually chorea gravidarum, a very severe form of rheumatic chorea in pregnancy a first attack is almost confined to primiparae under the age of 25

Its severity varies, from what may appear to be mere fidgetiness, to violent uncontrollable movements of the limbs, head and face, and trunk.

In an early and slight case the child is continually dropping things,

and all her movements are sudden and clumsy To elicit choreiform movements, ask her to hold both hands above her head after a few seconds involuntary movements appear in one or both hands or ask her to shake hands the strength of her grip continually varies, and it may be accompanied by a sudden jerking movement of the arm or ask her to put out her tongue, then put it back it is snapped back with startling rapidity Another characteristic is the attitude of the hands when the arms are outstretched they are held out with wrists markedly flexed and fingers hyperextended

When at all severe the nature of the movements is quite obvious. They are irregular in time and the same movement is not regularly repeated They cause considerable incoordination of voluntary movements objects are continually dropped or knocked over in trying to pick them up the child walks clumsily or in very severe cases may be scarcely able to walk, for what should be smoothly co-ordinated movements have become erratic. The swing of the arm in walking is apt to be lost, as in Parkinsonism.

She is seldom relaxed and quiet but is more or less incessantly disturbed by uncontrollable rapid jerky purposeless movements, of wrists, forearms, shoulders, head facial muscles, and sometimes of the trunk, in no regular sequence In many cases these movements are unilateral They usually cease during sleep Slight weakness of the limbs with hypotonia of the muscles, is invariable and it sometimes happens that choreiform movements on one side diminish and the arm and leg of that side become progressively weaker

The choreic child is emotionally unstable, and her speech may be affected. Choreiform movements rarely persist longer than about 2 months, but relapses are common, during which signs of endocarditis are especially apt to appear

Huntington's chorea

This is a rare hereditary disease, characterized by choreiform movements, and a slowly progressive mental deterioration, beginning in middle age.

Hysteria

Choreiform movements may occur in hysteria a clue to their nature being found in the cessation of movements when the patient thinks she is not under observation.

ATHETOSIS

In this disorder there are slow writhing involuntary movements, in the upper limbs more than the lower and when bilateral, in the face It

is due to a lesion of the old motor system. It may be congenital and bilateral. Unilateral athetosis is seen in infantile hemiplegia. Bilateral athetosis is very rare in adult life but unilateral movements can begin at any age—e.g. in the decline of life from a lesion of vascular origin.

These involuntary movements are so much increased by attempted voluntary movement as to interfere seriously with normal use of the limb. The movements disappear during sleep.

TICS

Habit spasm

The only disorder with which rheumatic chorea could reasonably be confused is habit spasm. This is the involuntary repetition of some natural movement: a shake of the head, a grimace, a shrug of the shoulder and so on in almost endless variety. Common in nervous children, they are increased by emotion, and cease during sleep. The feature that at once distinguishes habit spasm from chorea is the regular repetition of the same movement. It usually stops at about the age of 12 years.

Spasmodic torticollis

This is comparatively rare. It should be distinguished from congenital torticollis, a condition due to a growth defect of one sternomastoid.

Tonic or clonic spasms of neck muscles, which may be of organic or hysterical origin, pull the head into rotation, and often flexion to one side. If tonic, torticollis is sustained: if clonic, the head is repeatedly jerked into the abnormal position. Movements cease during sleep.

LIMPING IN CHILDREN AND ADOLESCENTS

A peculiar gait accompanies a large number of disorders of the locomotor system. Limping is the lame walk that results from pain, or weakness, or deformity or stiffness in one lower limb. It is not often the symptom that brings the patient for advice: rather will he complain of pain or other defect causing the limp.

The only exception to this is limping in children. It is common for a mother to bring her child for advice because of the appearance of a limp. The importance of it derives from the fact that limping is the earliest and often the only symptom of *tuberculosis of the hip*. The first step is to

determine if the limp originates in the hip joint or elsewhere The possibilities may be grouped

LIMP FROM A DISORDER OF THE HIP JOINT

Tuberculosis

Other disorders traumatic synovitis coxa vara congenital dislocation Perthes disease (Legg-Calvé-Perthes disease)

LIMP ORIGINATING ELSEWHERE

Spine tuberculosis (psoas abscess) [p 91]

Joints of the lower limb tarsus ankle knee tuberculosis [pp 119 154] Still's disease subacute rheumatism [pp 170 195]

Long bones chronic periostitis osteomyelitis sarcoma

Atrophic and spastic paresis

Minor disorders abrasions local infections chilblains minor injuries foot deformities inguinal lymphadenitis

THE HIP JOINT

Let us assume that the child has no pain, or admits to no more than a vague aching in the region of one hip and that a physical examination reveals no physical sign suggesting a condition in group II We then examine the hip joints with the greatest possible care.

Have the child undressed lying on a couch. Physical signs to be sought are

by inspection local swelling muscle wasting, especially of glutei deformity at the hip joint real or apparent shortening of the limb

by palpation local swelling tenderness altered position of the great trochanter

by testing passive movements compare the range of all movements at the hip joints on the two sides, testing in turn flexion, extension, abduction and adduction, internal and external rotation If all these movements on one side are restricted compared with the corresponding movements on the other side (and psoas abscess has been excluded) disease of the hip joint can be diagnosed with confidence *This must be assumed to be a tuberculous arthritis until the contrary is proved* as it may be by 1 X ray examination 2 complete recovery after a short period of rest

Confirmatory signs of arthritis are

deformity In the early stages the thigh is held slightly flexed, abducted and externally rotated at the hip (later it may be flexed adducted and internally rotated) This is not always easy to recognize at a glance Fully flex the sound hip so that the lumbar spine rests flat on the couch any flexion deformity at the other hip then becomes

obvious. The patient tries to bring the leg to lie flat on the couch. If he can do so no flexion deformity is present if he cannot, a rough measurement of the angle between the thigh and the couch when extension is checked gives approximately the amount of flexion deformity Rotation deformity is indicated by the direction of the foot. Abduction may be missed if the pelvis is tilted see that the anterior superior spines are level



FIG. 11
Tuberculosis of the hip joint in a child
(by kind permission of Dr Cecil Bull)

Muscle Wasting Wasting of the buttock muscles over the affected hip is seen early and is a very characteristic sign.

If these signs of arthritis are present, and X ray films of the hip joint show no abnormality and psoas abscess can be excluded the only alternative to a tuberculous arthritis is a traumatic synovitis. The distinction can only be made by observing the effect of some 2 weeks rest in bed. If after this time the signs are no longer found the condition is not tuberculous If they persist, it is assumed to be tuberculous, and X ray examination repeated at intervals.

Never omit examination of the lower dorsal and lumbar spine, for *psoas abscess* secondary to spinal tuberculosis, may closely mimic tuberculosis of the hip It forms a fluctuating swelling usually external to the femoral vessels. Caseous matter has extended downwards from a focus in the lower dorsal or lumbar spine along the psoas muscle, to reach a situation in front of the hip capsule The thigh is held flexed and rotated inwards or outwards, and all movements at the hip are restricted.

An X ray examination of the hip and of the lower dorsal and lumbar spine is carried out in all cases. At a very early stage the films of a

tuberculous hip will probably be normal Later there will be a marked osteoporosis a loss of definition of the bone trabeculae in the porotic area, and of the articular surface erosion of the articular surface though this is very variable in the time of its appearance [FIG 11]

The following conditions, which may be a source of confusion, can all be detected radiologically



FIG. 12
Perthes disease.

Epiphyseal Coxa Vara. This chiefly affects boys aged 10-17 years and both hips in about 25 per cent. of cases. A limp is the prominent symptom. At first adduction is of full range but later all movements may be restricted.

Congenital Dislocation of the Hip Girls are chiefly affected in one or both hips. The earliest symptom is a limp Unilateral cases are characterised by a marked shortening of the affected limb bilateral

cases by a considerable lordosis Abduction and external rotation limited, but other movements are of full range the femur can be mo up and down in its long axis

Perthes Disease The onset of this condition, a softening and f mentation of the femoral head usually begins between the ages years All movements at the affected hip may be restricted from mu spasm But quite early the X ray appearances are clear and cha teristic, for the femoral head is fragmented and deformed [FIG 12]

OEDEMA

OEDEMA OF THE ARM

Bilateral oedema of the upper limbs is usually caused by obstruc to the superior vena cava. Possible causes are thoracic aneur mediastinal growth, spread of thrombosis from the main venous tru

A commoner problem is the diagnosis of unilateral oedema of w the more important causes are

PAINFUL

Reflex sympathetic dystrophy [p. 83]
Thrombophlebitis axillary or subclavian
Aneurysm of the subclavian artery
Osteomyelitis of a long bone

PAINLESS

Lymphoedema
Angioneurotic oedema

Thrombophlebitis and aneurysm

Thrombophlebitis, axillary or subclavian is much less common t the corresponding affection of the iliofemoral vein in the lower li The onset is marked by pyrexia and malaise and at first the whole up limb is, moderately or severely painful The superficial veins are dila the skin cyanotic, and the whole limb is enlarged there may or may be pitting oedema. A tender thrombosed axillary vein can be felt.

Aneurysm of the axillary or subclavian artery may cause oedem the arm by pressing on and obstructing the vein It is rare

Osteomyelitis

Chronic osteomyelitis of humerus or radius [p 158] may recur intervals of months or years it may be, with no sinus formation. patient has recurrent attacks of aching in the bone, with oedema of surrounding soft tissues redness of the overlying skin, and tendern

associated with a mild malaise and pyrexia X ray films usually establish the diagnosis.

Lymphoedema

Lymphoedema results from obstruction of lymph vessels It affects the leg more commonly than the arm [pp 70 71]. At first the oedema is soft and pitting, like that of chronic venous insufficiency Later the skin and subcutaneous tissues become thickened and the limb is enlarged not only by oedema, but also by a general fibrosis of adipose tissue

Affecting one arm it must first be recognized as lymphoedema distinguished, that is, from the oedema of chronic venous insufficiency from angioneurotic oedema in which the swelling is transient and from causes of localized oedema

Then the cause, or the type of lymphoedema must be sought there are two broad clinical types—non-inflammatory and inflammatory

Non-inflammatory Lymphoedema In any case of lymphoedema, the first thing to do is to examine the lymph glands draining the limb with the possibility of malignant disease in mind The most important instances result from occlusion of lymph vessels by new growth in the axillary glands either metastases from carcinoma of the breast, lymphosarcoma, or Hodgkin s disease From a similar cause lymphoedema of an arm may follow after a very variable interval, removal of the breast and axillary lymph glands for carcinoma.

The non-inflammatory group includes the well known but uncommon Milroy s disease a congenital and hereditary lymphoedema of one leg, or one arm. It may be confused with congenital arteriovenous fistula but this causes increase in the length as well as the circumference of the affected limb

Inflammatory Lymphoedema Chronic lymphoedema may result from repeated attacks of acute cellulitis and lymphangitis High fever marks a sudden onset of swelling of the limb the part becoming red, hot and tender Fever quickly abates, inflammatory signs more slowly and some swelling persists. After a number of such attacks, permanent lymph oedema may be marked. Often the causal conditions are obscure In many cases it occurs spontaneously as do attacks of phlebitis. Others begin weeks or months after a local infection.

Angioneurotic Oedema. Angioneurotic oedema is often a familial condition. Because of its transient nature it is unlikely to cause difficulty in diagnosis. The patient is subject to attacks of oedema lasting a few hours to several days. The affected zone is restricted and may be in any part of the body Other symptoms are usually minimal but a few patients also suffer from abdominal pain and vomiting.

OEDEMA OF THE LEG

BILATERAL OEDEMA

There are many more possible causes of bilateral oedema of the lower than of the upper limbs. Most of them are problems in general medicine —the more important being acute and chronic nephritis, heart failure anaemia, cachexia starvation oedema beriberi drug oedema.

If these can be excluded, we are left with the following, which can be grouped according to the extent of the oedema

EXTENSIVE OEDEMA
- obstruction of the inferior vena cava
- some instances of lymphoedema

OEDEMA OF THE FOOT ANKLE AND LOWER LEG
- after prolonged bed rest
- a vasomotor disturbance in the elderly
- arteriosclerosis obliterans
- erythema induratum and chronic pernio
- disorders of the foot

Thrombophlebitis

Oedema of the whole of the lower limbs may be caused by obstruction of the inferior vena cava. If this is due to a spreading thrombosis, there will be a history of one leg affected first. Abdominal and pelvic examination will determine the presence or absence of pressure by new growth or a large ovarian cyst.

Lymphoedema

Lymphoedema is usually unilateral But both legs may be affected in lymphoedema *praecox (see next section)* Such cases must be distinguished from the fat legs of lipodystrophy Both affect women, and the appearance may be much the same Pitting is an unreliable guide, for it may be absent in both or present in both. The indurated skin and subcutaneous tissues of lymphoedema of long duration contrast with the more normal texture in lipodystrophy Seen during its development, the spread of lymphoedema proximally from the ankle distinguishes it from lipodystrophy which affects the whole limb from its first appearance.

Recognition of the cause of oedema confined to the feet, ankles and lower part of the leg should present little difficulty It is perhaps sometimes forgotten that it may be almost normal in the elderly who sit immobile for long periods and that it is fairly common in convalescence *after prolonged rest in bed Arteriosclerosis obliterans* is a possible cause. When associated with long-standing erythema induratum, or

with chronic pernio or with a local arthritic condition such as rheumatoid arthritis of the tarsal joints, diagnosis will turn on the nature of the nodules [p 44], or of the arthritis [p 119]

UNILATERAL OEDEMA

Causes of oedema of one lower limb are

THROMBOPHLEBITIS

LYMPHOEDEMA

lymphoedema praecox Milroy s disease lymphoedema from occlusion of lymph vessels by new growth inflammatory lymphoedema

OSTEOMYELITIS

ANGIONEUROTIC OEDEMA

LOCAL INFLAMMATORY CONDITIONS OF THE FOOT AND ANKLE

sprains, strains and injuries.

The oedema of lymphoedema is at first soft and pitting like that of chronic venous insufficiency later skin and subcutaneous tissues are indurated, and pitting is lost.

Thrombophlebitis

Thrombophlebitis of superficial veins does not as a rule cause oedema of the limb and when affecting deep veins of the calf oedema is rare. Obstruction of the popliteal vein causes only transient swelling of the lower part of the leg and the foot Iliofemoral thrombophlebitis, however brings a rapid and marked enlargement of the whole of the lower limb It has a sudden febrile onset with pain that may be quite severe The superficial veins are distended and the skin cyanosed there is tenderness in Scarpa s triangle A pitting oedema is obvious after the first few days.

Chronic venous insufficiency results from iliofemoral thrombophlebitis, or from repeated attacks in superficial veins. The limb is oedematous, the oedema subsiding after a night s rest in bed. The superficial veins are dilated and in cases of long duration the skin of the lower half of the leg is apt to be indurated and pigmented in this region eczematous patches and stasis ulcers develop.

Lymphoedema

This affects the leg more commonly than the arm [p 69] Two groups are distinguished

NON-INFLAMMATORY

Lymphoedema praecox occurs predominantly in girls and young women often the onset is at puberty In most cases one leg only is

involved a foot or ankle becomes oedematous, and usually the swelling gradually spreads to the whole of the lower limb The full development may take many months but the speed and extent of spread are somewhat variable

Oedema is at first relieved by elevation of the limb later only partially The skin becomes rough and indurated but does not show the changes of chronic venous insufficiency the subcutaneous tissues are firm and swollen. Pitting may not occur in cases of long duration. The condition is painless, but the limb feels uncomfortable and heavy

Lymphoedema may occur from occlusion of lymph vessels by metastases in lymph glands, from, for example, carcinoma of the uterus, vulva, prostate, and so on Swelling of the limb may be the first sign of new growth and should therefore always prompt careful examination with this possibility in mind. Hodgkin s disease and lymphosarcoma are sometimes responsible.

In this group of non inflammatory lymphoedema comes Milroy s disease, a congenital and hereditary condition. One leg—or one arm—is larger than its fellow at birth. The limb shows a pitting oedema, *considerably reduced after elevation.*

INFLAMMATORY

In contrast to this slowly progressing swelling of the limb are instances of inflammatory lymphoedema, in which the patient has one attack after another of cellulitis and lymphangitis, each leaving the limb more swollen than before Each attack is of sudden onset and accompanied by severe febrile symptoms.

If we are dealing with neither thrombophlebitis nor lymphoedema, then probably some local inflammatory or traumatic condition is responsible. The familial angioneurotic oedema is a possibility attacks are recurrent and transient. Activation of a chronic osteomyelitis is accompanied by local oedema. Or oedema may be due to a local disorder in the foot or ankle e.g. rheumatoid arthritis, osteoarthritis, gout, chronic sprain of the ankle joint, pes valgus.

PAIN IN THE ARM

[See also Pain in the Hand, p. 142 and Pain in the Shoulder p. 171]

The disorder will be in one of these four categories

LOCAL LESIONS IN THE ARM

PAIN REFERRED FROM THE SHOULDER GIRDLE

BRACHIAL NEURALGIA

REFLEX SYMPATHETIC DYSTROPHY

LOCAL LESIONS

Arthritis, bone diseases and so on—are usually recognized without difficulty Pain from sudden arterial occlusion may be mentioned here it can occur in the arm, but is much more common in the lower limb [p 43]. It is accompanied by coldness, pallor and numbness of the hand Pain does not always come on abruptly The diagnostic sign of most importance is absence of arterial pulsation the brachial radial and ulnar pulses are examined Arterial pulsation should be examined as a routine in all cases of pain in a limb accompanied by coldness, numbness and tingling.

REFERRED PAIN IN THE ARM

This is possible from any painful condition in the neck and shoulder Features distinguishing referred pain from brachial neuralgia are these

1 Referred pain in the arm is less severe than that felt more locally in the neck and shoulder the reverse is true for brachial neuralgia which is most intense in the arm.

2. Pain in the arm associated with a lesion in the cervical spine is much more likely to be root pain than referred pain referred pain from the cervical spine is felt chiefly if not entirely in the shoulder girdle region.

3 Neuralgia is a paroxysmal pain characteristic are bouts of throbbing pain in the length of the limb Referred pain is more constant.

4 Neuralgia is almost always associated with paraesthesia in the hand referred pain never

BRACHIAL NEURALGIA

The causes are

ROOT PAIN

cervical disc protrusion
cervical spondylosis
disease of the vertebrae tuberculosis osteomyelitis new growth ankylosing spondylitis
disease of the cord or meninges cervical cord tumour pachymeningitis
herpes zoster
carcinoma of apex of lung

PRESSURE ON NERVE TRUNKS

cervical rib syndrome
costoclavicular syndrome
pressure by aneurysm of the subclavian artery
pressure by tumour in the posterior triangle

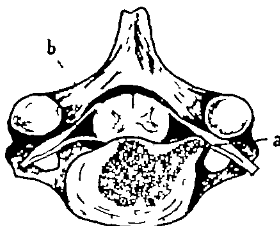


FIG. 13
Diagram of cervical disc protrusion
(*a*) lateral protrusion compressing nerve root
(*b*) central protrusion compressing cord.

ROOT PAIN

Cervical disc protrusion

The type case is a middle-aged patient, in whom pain in the arm came on with, or a few days after a painful stiff neck, which may or may not have subsided. Less severe pain is felt in the zones of the upper trapezius, supraspinatus and pectoral muscles, and the vertebral border of the scapula Severe paroxysms of pain in the length of the limb are provoked

by movements of the arm or neck, or by coughing. Pain at night may be intense, and is usually much worse than during the day Prominent also are paraesthesiae in some of the fingers.

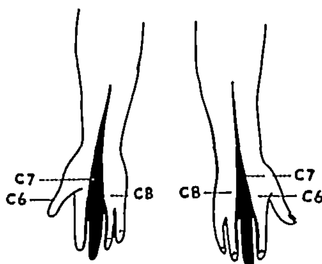


FIG. 14
Approximate area of dermatomes C.6 and C.7 in the hand.

This is the clinical picture of root pain the commonest cause is cervical disc protrusion [FIG 13]. The probability is increased if on examination we find the normal cervical curve flattened the neck tilted to one side its movements restricted by muscle spasm, especially extension, and side-bending to the side of the painful limb

There are now two further inquiries which are quite essential by them the diagnosis will be confirmed or refuted.

I EVIDENCE OF INVOLVEMENT OF C.6 OR C.7 NERVE ROOT

Distribution of Pain. A patient with a lesion of C.6 nerve root feels pain most intensely in the anterolateral part of the upper arm, stopping at the elbow C.7 root lesions give pain in the posterolateral part of the upper arm and on the extensor side of the forearm

Paraesthesia. Tingling and numbness in the thumb and index are typical of C.6 lesions in the index and middle fingers of C.7 [FIG 14]

Muscular Weakness (and wasting in later stages of severe attacks) In compression of C.6 root some weakness may be found in the biceps In compression of C.7 root it is most easily shown in the triceps [Table p 12]

Tendon Jerks A C 6 lesion is associated with loss or diminution of the biceps jerk a C 7 lesion with loss or diminution of the triceps jerk

Sensory Loss This is always very slight a diminution of pain sensation on part of the thumb and index in C.6 lesions, and on part of the index and middle fingers in C.7

The more important of these signs are summarized in the Table.

	C.6 (C.5-6 DISC)	C.7 (C.6-7 DISC)
Distribution of pain	anterolateral upper arm	posterolateral upper arm extensor forearm
Paraesthesia	thumb and index	index and middle
Tendon jerks	loss of biceps jerk	loss of triceps jerk
Muscular weakness	biceps	triceps

Practically all cervical disc protrusions giving root pain are from the discs C.5-6 or C.6-7 compressing either C 6 or C.7 roots respectively The diagnostic power of their localization will be obvious when considering some of the other causes of brachial neuralgia.

2. X RAY FILMS OF THE CERVICAL SPINE

With a small protrusion, X ray appearances may be quite normal. Often some narrowing of the affected disc space can be seen Another suggestive feature is absence of the normal cervical curve, the vertebral bodies standing up as an erect column

If in lateral or oblique views small posterior osteophytes can be seen the lesion is probably an old one for this represents a local bone reaction to the protrusion and probably takes at least a month to develop Anterolateral lipping, and marginal sclerosis of adjacent vertebrae indicate disc degeneration

With evidence of root pain due to a lesion of C.6 or C.7 and X ray films either normal or showing only some narrowing of the C.5-6 or C.6-7 disc space, the diagnosis is confirmed with two possible sources of error

1 a secondary deposit of carcinoma so early that it gives no radiological evidence of its presence

2. a cervical cord tumour or pachymeningitis.

Metastatic carcinoma [p 93] is not excluded even if X ray films are normal, when they have been obtained soon after the onset of pain. If there is known to be, or to have been, a primary growth, the films must be repeated after a short interval

Cervical cord tumour

Extramedullary tumours are neurofibromata, arising from nerve

roots and meningiomata. Intramedullary tumours, much less common are usually gliomata.

Root pain is the early symptom of both, and the clinical picture may closely resemble that of disc protrusion The chief differences are 1 in the course of the disorders severe root pain from a disc subsiding as a rule after a few weeks, from a tumour continuing for months 2 in the appearance sooner or later in cord tumours of signs indicating involvement of more than one root, and pyramidal signs in the legs indicating cord compression The cord however may be compressed by a centrally placed disc protrusion and distinction clinically may scarcely be possible. In any event, cord compression will indicate operation which will establish the diagnosis.

Selective muscular weakness and wasting in cord tumours, of segmental distribution, may be well marked. C.5 is the main root supply of the supraspinatus, infraspinatus and deltoid C.8 of the flexors of the wrist and fingers T 1 of the intrinsic muscles of the hand. Root pain muscular weakness and wasting in the arm or hand, and pyramidal signs in the legs, is the late picture of cervical cord tumour

Syphilitic pachymeningitis is a localized thickening of the meninges in the cervical part of the cord which may cause root pain in the arm with paresis. Ultimately there are signs of cord compression.

Herpes zoster is occasionally confusing for a day or two for pain precedes the rash, which usually appears about the 4th day The arm is much less commonly affected than is the trunk but any root of the brachial plexus may be involved Serious and intractable neuralgia may persist in elderly patients.

Disease of the vertebrae

Root pain from disease and collapse of a vertebra may closely simulate the picture of cervical disc protrusion but the condition is revealed by X ray films of the cervical spine.

X ray appearances of metastatic carcinoma may be seen when there has been no indication of a primary growth Here may be mentioned Pancoast's tumour—carcinoma of the apex of the lung. Radiographs may show it invading the bone in the region of T 1 root, or T 1 and T.2 so it causes, in addition to severe shoulder pain, pain down the medial side of the arm and wasting of the small muscles of the hand On the same side will be found a small pupil and ptosis, from involvement of the stellate ganglion.

Tuberculosis of a Cervical Vertebra Mainly a disease of children, some 12 per cent. of all cases occur in adults. Of the several regions of the spine it occurs least often in the neck. There is a dull continual pain

in the neck the head is held very still for pain is intensified by attempted movement. The patient walks gingerly to avoid jars to the spine. The triad local pain, tenderness, and rigidity which in children at once arouses suspicion of spinal tuberculosis, is well marked.

The earliest X ray sign is the appearance of bone rarefaction and absorption near the superior or inferior surface of a vertebral body resembling a Schmorl nodule Or it may appear as a localized erosion of the adjacent surfaces of two vertebral bodies. Later rarefaction becomes extensive, with little or no sign of bone regeneration. The outline of the body becomes eroded and the body is gradually compressed to a wedge shape The infection may spread to neighbouring vertebrae An abscess is likely to form early and shows in a lateral radiograph by a displacement forwards of the pharynx (retropharyngeal abscess) it shows very clearly if the contents become calcified

Root pain may be bilateral as it may in other diseases of the vertebrae.

Osteomyelitis of a Vertebra. This condition is rare and very rare in the neck. But it does occur and its distinction from tuberculosis or metastatic carcinoma may for a time be quite difficult. The infected body appears in radiographs collapsed oedema, or effusion into the retropharyngeal tissues, gives the appearance of an abscess. But the course of the illness is different there is a febrile onset rapid relief of pain after antibiotic treatment and subsequently rapid and uninterrupted bone regeneration, contrasting with the usual very slow healing of a tuberculous process [FIG 15].

Cervical Spondylosis [p 172] Radiological evidence of cervical disc degeneration, in any space from C.3-4 down, but especially in C.5-6-7 is so common and so often symptomless, that its association with a syndrome cannot necessarily be taken as a causal association.

Cervical spondylosis is now generally regarded as responsible for a number of syndromes, one of which is root pain. This could arise 1 from simple narrowing of the foramen 2. from the encroachment of osteophytes into the foramen—and this especially when secondary osteo-arthritic changes have occurred in the facets 3 from root-sleeve fibrosis —a fibrotic reaction in the dural sleeve of the nerve roots, which is said to occur not only at the level of the degenerate discs, but also above and below

Whatever the cause of the root irritation, it is clear that the symptoms are likely to be multiradicular The pain is more diffuse and far less well localized than that of disc protrusion. It is usually less severe, and may be intermittent. It is sometimes bilateral though more severe on one side than the other Paraesthesia often affects all the fingers of the



(*a*)



(*b*)

FIG. 15

Osteomyelitis of a cervical vertebra.
(*a*) Film 7-8 weeks after the onset of symptoms destruction and collapse of C.5 considerable enlargement of the retropharyngeal soft tissues suggesting a large abscess.
(*b*) Film 8 months after (*a*) bone regeneration has occurred the retropharyngeal effusion has disappeared

painful limb Motor signs, if present, are likely to indicate a multi-radicular origin

With this picture, and clear X ray evidence of cervical disc degeneration cervical spondylosis is the probable diagnosis. But it is wise to think for a moment that we may be mistaken.

Ankylosing Spondylitis [p 87] I recall the case of a woman aged 60, in which restriction of neck movements was severe, and X ray changes

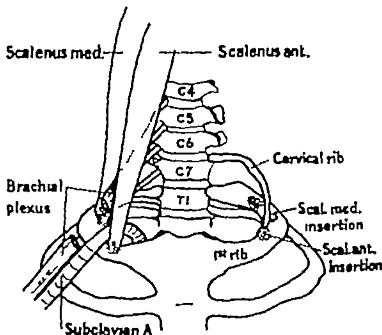


FIG. 16

The left of the diagram shows the subclavian artery and brachial plexus passing through the space bounded by the scalenus anterior and medius and first rib the right shows how this space may be encroached on by a cervical rib or its fibrous prolongation to the first rib.

of cervical disc degeneration only very moderate. She had no pain or stiffness in any other part of the spine but the disparity between the marked stiffness and slight disc degeneration suggested that this might be one of those unusual instances of ankylosing spondylitis in which symptoms and signs first appear in the neck. Radiographs of the sacro-iliac joints showed unmistakably the very characteristic changes of bilateral sacro-iliitis and the E.S R. was considerably increased.

PRESSURE ON NERVE TRUNKS

There are some causes of brachial neuralgia that may not be thought of and may therefore be missed If the X ray diagnosis of cervical spondylosis is accepted too readily

I remember particularly a man with a rapidly growing lymphosarcoma, affecting glands of the posterior triangle of the neck and of the axilla who presented as brachial neuralgia the correct diagnosis was not made for some few weeks, when the glandular swelling became obvious.

Pressure from an aneurysm of the subclavian artery is very rare but will not be missed if routine examination of the posterior triangle and of the axilla is always made in brachial neuralgia.

The common causes of pressure on nerve trunks of the brachial plexus are

1 An abnormal bridge formed by a cervical rib and its fibrous prolongation to the scal ne tubercle of the first rib The lowest trunk of the plexus and the subclavian artery are subject to pressure as they cross over it *cervical rib syndrome* [FIG 16]

2. Compression between the scalenus anticus muscle in a state of spasm, and tendinous fibres of the lower medial edge of the scalenus medius muscle *scalenus syndrome*

3 Fatigue and loss of tone of the shoulder girdle muscles permitting intermittent pressure between the clavicle and the first rib *costoclavicular syndrome*

Cervical rib syndrome

A cervical rib is infrequent it is present in about 0 06 per cent of the population It is a congenital abnormality but never causes symptoms until adult life many possibly 50 per cent., never do so

Diagnosis rests on a characteristic syndrome—not on the radiological demonstration of a cervical rib [FIG 17] Sometimes when presented with this picture we find no cervical rib in an X ray film such cases are very probably instances of the scalenus syndrome.

The type case is a young woman complaining of *pain and paraesthesia in one arm*, often of some months duration. Pain is felt either along the ulnar side of the forearm and hand with tingling in the ring and little fingers or less commonly along the radial side with tingling in the thumb and index It is aggravated by carrying weights or by the use of the arm at work. It is particularly bad at night but relief can often be obtained by elevating the arm.

Vasomotor symptoms in the painful forearm and hand are fairly common attacks of prolonged cyanosis and coldness or attacks of blanching of the fingers.

These are the presenting symptoms later muscular atrophy may develop Ulnar-sided pain is associated with wasting of the hypothenar eminence and the interossei—and so a partial claw hand radial-sided

pain with wasting of the lateral part of the thenar eminence. There may also be found a winged scapula on the affected side, from paresis of the serratus anterior due presumably to stretching of the long thoracic nerve.



FIG. 17
Rudimentary cervical rib often connected by a fibrous band to the scalene tubercle of the first rib

Peculiarities distinguishing this syndrome from root pain due to spondylosis are

1 Pain is most severe in, and is almost confined to the forearm and hand this is not so in spondylosis.

2 Paraesthesia affects either the ulnar or the radial side of the hand all the fingers are affected in spondylosis.

3 Relief is obtained by elevating the arm this is not so in spondylosis

4 Vasomotor symptoms are common they are very rare in spondylosis.

A helpful diagnostic manoeuvre is to ask the patient to rotate the head to the affected side, and raise the chin, at the same time inspiring. If the radial pulse is not obliterated a diagnosis of cervical rib or scalenus syndrome is improbable

Because of paraesthesia, and wasting of the hypothenar or thenar

eminence the cervical rib syndrome may be confused with ulnar or median nerve lesions Ulnar paralysis [p 12] is, however almost painless. Compression of the median nerve in the carpal tunnel is a cause of acroparaesthesia [p. 187] nocturnal attacks of painful tingling in the fingers. The chief difference is that pain and paraesthesia and sensory loss have a strictly median distribution this is not so in the cervical rib syndrome.

The costoclavicular syndrome is discussed under acroparaesthesia [p 186]

REFLEX SYMPATHETIC DYSTROPHY

A number of painful disorders of the upper limb associated with vasomotor and trophic changes in the hand probably have the same mechanism and have been called reflex sympathetic dystrophy The nature of the disturbance is not understood that it is essentially an autonomic disturbance is suggested by the success of treatment by sympathetic block, or sympathectomy

Among these disorders are

Causalgia
Sudeck s atrophy (post traumatic osteoporosis)
The shoulder-hand syndrome
idiopathic following coronary infarction post hemiplegic post herpetic associated with severe cervical spondylosis

Causalgia

This is an occasional sequel to injury of a peripheral nerve—usually the median nerve. It is characterized by 1 very severe continuous throbbing burning pain in the limb 2. hyperaesthesia 3 extensive trophic changes vasomotor disturbances.

Sudeck s atrophy

Sudeck, a surgeon of Hamburg, first described these remarkable cases in 1900 Soon after an injury in the region of the wrist (or less commonly the ankle) which may have been quite trivial the patient experiences burning pain in the injured part, which gradually extends until it involves the whole limb It increases in severity some patients after a few weeks enduring such agonizing paroxysms that they flinch from any examination. It is associated at first with oedema, and with a moist cyanotic skin. The hand is held pronated immobile. X ray films show an intense osteoporosis of the hand and carpus, and perhaps of the lower end of the radius and ulna. Later oedema may subside the

limb atrophies the skin remains cold, moist and cyanotic, and torment ing pain persists.

Spontaneous arrest may occur after 2 to 3 months, but often the disorder lasts a year or longer

Pain oedema and osteoporosis in the region of the wrist may suggest acute arthritis gout tuberculosis of the carpus or tenosynovitis. The far greater severity and extent of the symptoms and signs in Sudeck s atrophy should be a sufficient criterion for distinction Leriche has pointed out that whereas plaster immobilization eases the pain of tuberculosis, pain continues with all its severity in Sudeck s atrophy

The shoulder-hand syndrome

The first stage lasting 3 to 6 months, is characterized by pain and restricted movement in the shoulder—much the same symptoms as those of acute subacromial bursitis—followed by swelling, pain and stiffness of the hand and fingers. The skin of the hand is smooth and taut, at first dusky pink or red later pale and cyanotic. The fingers are held in slight flexion and their movement is restricted. X ray films show a slight osteoporosis of hand and shoulder regions

The second stage also lasting 3 to 6 months is one of gradual relief of shoulder and hand pain and swelling but stiffness and flexion deformity of the fingers become more pronounced. The subcutaneous tissues and the intrinsic muscles atrophy and often there are early signs of Dupuytren s contracture Osteoporosis becomes more marked

The third stage is indefinite in duration Atrophy of the hand increases. Osteoporosis of the hand and humeral head becomes very marked. Trophic changes increase

Probably there are incomplete forms of this syndrome hand symptoms only—or only the shoulder affected The full syndrome should present little difficulty in diagnosis If the hand only is affected, the condition resembles Sudeck s atrophy if the shoulder it is indistinguishable from acute subacromial bursitis.

PAIN IN THE BACK

The minimum data needed for diagnosis of any case of pain in the back are those provided by 1 the history 2 routine clinical examination 3 clinical examination of the spine 4 radiography of the spine, and—in low back pain—of the pelvic bones and upper third of the femur In some instances special investigations are necessary

The probability is that pain is referred from the spinal column But when no spinal lesion can be demonstrated there are 4 possibilities

1 there is a spinal lesion though we cannot demonstrate it
2 pain is referred from a soft tissue lesion
3 pain is referred from visceral disease
4 there is a cause other than referred pain this includes the ill-defined condition called fibrositis

LOW BACK PAIN

The chief causes of low back pain are

SOMATIC

REFERRED PAIN FROM THE SPINAL COLUMN

Common disc protrusion disc degeneration osteo-arthritis of the apophyseal joints senile osteoporosis

Relatively uncommon ankylosing spondylitis Paget s disease tuberculosis metastatic carcinoma myeloma osteomyelitis

REFERRED PAIN FROM A SOFT TISSUE LESION

Myofascial or ligamentous sprain chronic strain from postural errors, lumbar scoliosis, spondylolisthesis, and possibly minor congenital defects chronic sacro-iliac strain

ACUTE MYOSITIS

FIBROSITIS

VISCERAL

UPPER LUMBAR REGION—REFERRED PAIN FROM

Kidney disease renal calculus chronic pyelonephritis malignant growth of the kidney perinephric abscess

Peptic ulcer especially invading the pancreas carcinoma of the pancreas

SACRAL—REFERRED PAIN FROM PELVIC DISEASE

Chronic salpingitis carcinoma of the uterus uterine fibroid retroversion of the uterus uterine prolapse ovarian tumour carcinoma of the prostate carcinoma of the rectum ischiorectal abscess spastic constipation

PSYCHOGENIC

HYSTERIA

Data from the history are often of greater diagnostic value than the clinical examination. Inquire therefore in some detail about the characters of the pain and about symptoms referable to the abdominal

organs, including the urinary tract, uterine and bowel functions and about the many symptoms of anxiety neurosis. When we have finished taking the history we should know whether the pain is probably somatic or probably visceral in origin.

The examination is guided by the probability that pain is referred from the spine. We examine the spine, noting abnormalities of posture and movements, and tenderness over the vertebrae and we get X ray films of the lumbar spine and pelvic bones in all instances. As a routine we examine the abdomen Pelvic and rectal examination is only necessary 1 when there are pelvic symptoms 2. when no adequate explanation is found by the routine data.

PAIN REFERRED FROM THE SPINAL COLUMN

There are some who would assert that, if we have excluded the visceral and psychogenic groups, practically all lumbago is referred somatic pain, and that the operative lesion is always in the spinal column I am sure it is not so But some abnormality in the spinal column will be found in I should say well over 50 per cent. of instances.

Posterior protrusion of an intervertebral disc

At times the history of low back pain is almost pathognomonic. During some everyday action—often it is rising from a stooping position —a man is stricken with crippling lumbar pain of alarming severity He can stand only with knees bent, for his lumbar spine is flexed and pulled over to one side in an agony of painful spasm. He is immobilized for a minute or two and can move only with immense effort.

I know of only two causes for this paroxysm sudden collapse of a vertebral body from some disease of the bone, and protrusion of an intervertebral disc. A history of a succession of these seizures over a number of years is almost diagnostic of disc protrusion.

Disc protrusion is rarely purely traumatic. It is preceded by some *morbid process* that weakens the annulus, *allowing protrusion* of the nucleus on very slight exertion [FIG 18] and this morbid process may or may not be symptomless. So an acute attack may be the only event or it may be preceded or followed by many months of backache.

The same crippling pain with locked spine may be ingravescent reaching its full intensity only after 24 hours or longer but ultimately the picture is typical

Lumbar signs in acute disc protrusion are characteristic. The normal curve is flattened, and the spine lists to one side it is held by muscle spasm rigid in this position of sciatic scoliosis As pain lessens some movement returns but lateral flexion to the side that would correct the

list is promptly checked by severe pain Some clear up completely but some leave a chronic low backache and then diagnostic signs disappear

X-ray appearances of the lumbar spine may be normal or in the lateral view the L.4-5 or L.5-S 1 disc space may appear narrow and not normally wedged The L.4-5 space is normally slightly wider than L.3-4 if it appears narrower the change is probably significant. The width of the L.5-S 1 space is so variable in health that no significance can be attached merely to an unusually narrow space Local bone reaction indicates an old lesion.

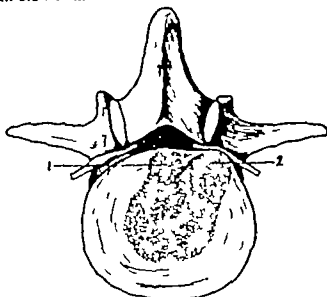


FIG 18
Diagram of lumbar disc protrusion. 1 central protrusion that, if large enough, may compress the cauda equina 2. lateral protrusion compressing a nerve root.

Be especially careful in adolescents and young adults It is then that ankylosing spondylitis begins, diagnosable by the X ray appearances of the sacro-iliac joints and in no other way It is then that tuberculosis of a sacro-iliac joint, rare though it is, may occur

Be especially careful also in patients who have had an operation for carcinoma. Bone may be affected by metastatic carcinoma and be radiologically normal for a time Severe lumbar pain in these cases is probably due to metastases if necessary repeat the X ray films after an interval

Ankylosing spondylitis (RHIZOMELIC SPONDYLITIS OF PIERRE MARIE)

Some 90 per cent. of patients with this disease are men. The earliest manifestations are likely to appear in adolescence Common modes of onset are

1 Transient pains in the limbs or joints, without physical signs, lasting weeks or months, and recurring after long periods of freedom.

2. An arthritis of the rheumatoid type affecting one of the limb joints.

3 Sciatica.

4 Lumbosacral pain

There are rare instances in which the first signs appear in the cervical spine

When seen at an early pre-spondylitic stage, diagnostic errors are very common Fortunately there is a diagnostic sign by which they can be avoided characteristic radiological changes in the sacro-iliac joints. X ray examination of the sacro-iliac joints must be carried out on all patients betwen the ages 14-25 complaining of recurrent limb pains, of sciatic neuralgia, of lumbosacral pain or of an arthritis of the rheumatoid type affecting one of the larger limb joints.

The characteristic changes are a loss of definition of the fissures, the line being broken by irregular zones of rarefaction indicating erosion later the fissures are no longer seen, their place being taken by irregular rounded zones of rarefied bone linearly arranged (a rosary effect) and there is sclerosis of the bone adjacent to these rarefied areas. This *bilateral sacro-iliitis always ends—probably after some 5-7 years—in* complete bony ankylosis [FIG 19]

Bilateral sacro-iliitis occurs in Reiter s disease [p. 207] it is not yet known in what percentage ankylosing spondylitis develops.

The phase of progressive spinal rigidity usually begins with pain and stiffness in the lumbar region

The type case is a young man usually under 35 and often 20-25, who gives a history of having suffered in adolescence from one of the pre-spondylitic symptoms mentioned above. He now complains of lumbar pain and stiffness and on examination, a flattening of the lumbar curve is noted, and a very great diminution of movement in this part of the spine The diagnosis is established beyond all doubt by the X ray appearances of the sacro-iliac joints Films of the lumbar spine may be normal or they may show the beginnings of the changes described below The E.S R is almost always raised

Over a number of years pain and stiffness of the spine ascend to the dorsal and ultimately the cervical regions. When the morbid process reaches the dorsal spine the ribs become ankylosed to the vertebrae, and respiratory thoracic movements are no longer possible. And at this stage the sufferer is apt to become more and more round shouldered and stooping, from an increasing kyphosis in the upper dorsal region, while the lumbar and lower half of the dorsal spine are almost in a straight line



(*a*)



(*b*)

FIG. 19

The sacro-iliitis (bilateral arthritis of the sacro-iliac joint) associated with ankylosing spondylitis (*a*) active stage—note fissures ill-defined sclerosis of adjacent bone (*b*) complete ankylosis—the fissures have disappeared.

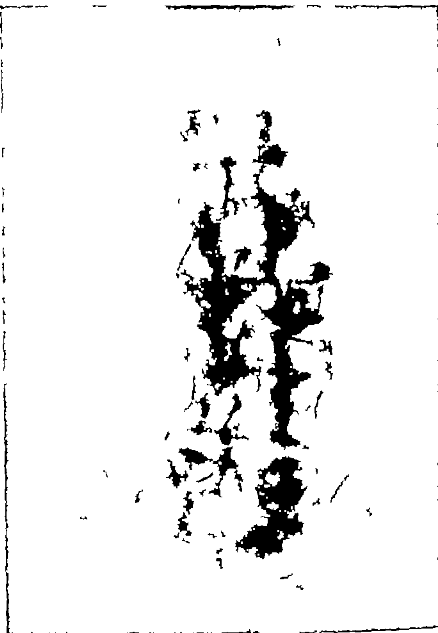


FIG. 20

Ankylosing spondylitis—lumbar spine. Note decalcification of vertebral bodies bamboo appearance from calcification of the edges of the intervertebral discs ankylosis of the sacro-iliac joints.

In the fully developed poker back radiological changes are highly characteristic ankylosed sacro-iliac joints extreme osteoporosis causing excessively bi-convex intervertebral discs calcification or ossification of spinal ligaments bony ankylosis of the apophyseal and costo-vertebral joints [FIG 20]

Disease of the Eye in Ankylosing Spondylitis Uveitis occurs sufficiently often in ankylosing spondylitis to be of some diagnostic value. In one series of 200 cases it was found in 8 5 per cent Attacks may occur early in the disease Uveitis in a young man therefore at once suggests the need for X ray examination of the sacro-iliac joints It varies in severity There may be slight pain in the eye, some photophobia and a sluggish pupil reaction—symptoms of a mild iritis which abates in 8-10 days. The iritis may be more marked with peri-corneal injection Photophobia may be severe, and rarely there may be severe pain Attacks may clear with no residual signs but in the most severe cases posterior synechiae may persist and cause impaired vision.

Diseases with which ankylosing spondylitis may be confused are

1 In the pre-spondylitic stage juvenile rheumatism joint tuberculosis lumbar disc protrusion

2. In the stage of progressive spinal rigidity adolescent kyphosis other causes of spinal osteoporosis

Tuberculosis of the spine

More than 80 per cent. of all cases of spinal tuberculosis begin before the age of 10 years but the disease can begin at any time in adult life. About a quarter of all cases are in the lumbar spine.

In children the triad local pain, widespread spinal rigidity and local tenderness, so strongly suggest tuberculosis as to be enough for a provisional diagnosis. Sometimes a psoas abscess is an early sign, causing a bulge in the groin and a painful limp necrotic material tracks down the psoas sheath and beneath the iliacus fascia passing under the inguinal ligament and pointing in the thigh lateral to the femoral vessels.

In adults diagnosis rests on the radiological appearances. The earliest change is an erosion of one of the surfaces of a vertebral body or of the adjacent surfaces of two vertebrae, which at first sight could be confused with a Schmorl node The bone however is clearly eroded in contrast to the sclerosed clean wall of the depression that is a Schmorl's node. In the little area of bone destruction a small sequestrum may be seen. The area of bone immediately around the erosion is rarefied there may be slight sclerosis outside this, but usually there is none [FIG 21] (This patient was a woman aged 39 who gave a history strongly



(a)



(b)

FIG. 21

Early tuberculosis of the spine (a) There has been a protrusion of the disc L.3–4 erosion of the adjacent vertebral bodies, especially of the lower surface of L.3 has occurred (b) lateral tomogram showing extensive erosion of the lower surface of L 3

suggesting lumbar disc protrusion two years previously with residual backache A film of the chest taken later showed appearances of active tuberculosis) The affected body becomes more rarefied and gradually compressed. Later the infection may spread to bodies above and below which become rarefied and eroded In the lumbar region a para vertebral abscess, in the region of the psoas may not show until it is calcified

Tuberculosis of a sacro-iliac joint is described under Pain in the Hip [p 138]

Osteomyelitis of a vertebral body

Staphylococcus aureus infection of the vertebrae is very uncommon but it does occur and may be mistaken for tuberculous infection, or a metastatic deposit. It may begin as an insidious febrile illness, with pain in the back and local tenderness. Most of the cases reported have been in the lumbar region X ray films may at first be normal, and it may not be possible to make a diagnosis immediately Later the infected vertebral body appears rarefied, and there is some bone destruction in which sequestra may be seen. The body may collapse If healing occurs, successive films will show a rapidly increasing sclerosis—far more rapid than in the healing of a tuberculous infection

* * * * * *

Primary tumours in the spine are rare. Sarcoma very rarely affects the vertebrae. Myelomatosis is a rare disease, but it involves the vertebrae in 50-70 per cent. of cases.

Metastatic carcinoma

Metastatic carcinoma is the commonest malignant tumour in the spine. The primary growth may be in these organs, in order of decreasing probability prostate (40 per cent.) breast (30 per cent.) kidney gastro-intestinal tract female genital organs thyroid lung. Evidence of secondary deposits in bone may be obtained before there is any indication of a primary growth carcinoma of the prostate, for instance may be giving no symptoms when metastases in bone have appeared

The first symptom is local pain and tenderness in the spine With an osteolytic growth the affected vertebra rarefies and collapses root pain of great severity may follow With an osteoplastic deposit the body does not collapse, and pain as a rule is less severe. Bone may contain quite large metastases and be radiologically normal for a time Patients who are known to have or to have had, a primary carcinoma, and who develop localized pain and restricted movement in the spine probably

have a metastatic deposit even with negative X ray films fresh films should be obtained after a short interval

The X ray appearances of an osteolytic metastasis are severe osteoporosis of a vertebral body and probably collapse Early rounded areas of porosis in the vertebral body may be seen if several bodies are so affected confusion with myelomatosis is possible Later the whole body becomes completely decalcified, and lateral views show it to be

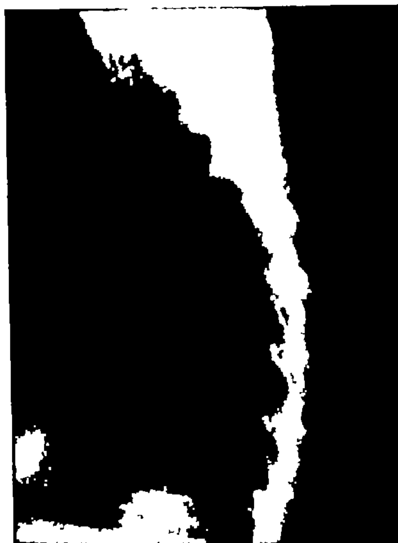


FIG. 23a
Myelomatosis dorsal spine showing general rarefaction, and collapse of the body of T 6.

growths in the bone marrow and metastatic deposits. It affects males more often than females, in late middle age The marrow of any bone may be affected most common are vertebrae ribs, sternum and skull.

Early diagnosis is the difficulty when multiple osteolytic lesions are seen in radiographs of the vertebrae, the ribs and the skull, diagnosis is comparatively easy but a late stage of the disease has arrived This is the classical textbook description. The presenting symptom is pain in the back, and X ray films of the spine show multiple rounded osteoporotic areas in the vertebrae, called punched out because there is no

local bone reaction, and their boundaries are sharp There may be collapse of one or more vertebral bodies.

Difficulty is found

1 When the beginning of the disease is a solitary multilocular cystic tumour in the spine, a rib the manubrium, or another bone. Such an isolated lesion may cause destruction of a vertebral body which may radiologically be confused with secondary carcinoma.

2. When X ray films show no more than a general porosis of the spine, with perhaps collapse of one or more vertebral bodies [FIG. 23a]

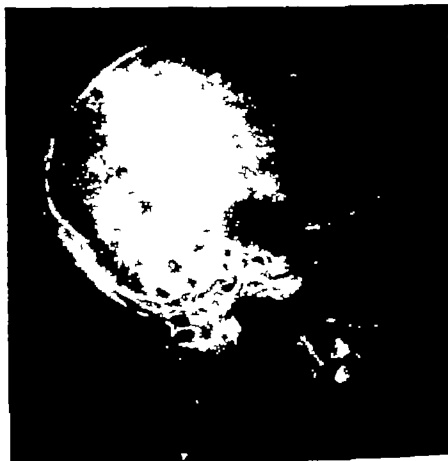


FIG. 23b
Myelomatosis skull.

In both forms the common presenting symptom is local pain, often persistent backache Both forms are followed eventually by typical multiple osteolytic lesions The solitary myeloma occurs at an average age of 40 years, the diffuse form over 60

Biopsy of a solitary tumour may be practicable it will show histologically a typical plasmocytoma If not possible and in all cases of diffuse decalcification of the spine—especially in cases of backache in the elderly with radiological osteoporosis, unrelieved by rest—the special investigations for myelomatosis should be carried out These are

1 X ray of skull and ribs in addition to vertebrae They may show typical circumscribed osteolytic lesions [FIG 23b]

2. Blood count and E.S R There is a greatly increased E.S R. and some degree of anaemia

3. Total serum protein albumin and globulin There is a rise in total protein globulin is increased with reversal of albumin/globulin ratio

4 Urine for Bence Jones proteose a precipitate forms on warming the urine to 50° C. disappears on boiling, and reappears on cooling. Its absence is of no significance it is negative in solitary myeloma, and positive in 50 per cent of multiple cases.

5 Practically all cases of myelomatosis show an abnormal protein by electrophoresis, either in the plasma, or in the urine

6. Sternal puncture may demonstrate typical myeloma cells

Bone tumours in the region of the hip

Reference may be made here to recorded instances of sarcoma in the neck of a femur secondary hypernephroma in the acetabulum and secondary carcinoma in the ischium giving lumbar and hip pain, radiating to the leg, and treated for a time as intervertebral disc lesions.[1] It is wise therefore in all doubtful cases to X ray the proximal part of the femur

* * * * * *

The other diseases of the spine cause as a rule chronic backache rather than severe pain They are spondylosis, spinal osteoporosis, and Paget s disease. All are readily demonstrated by X ray films.

Spondylosis

Disc Degeneration and Osteophytosis of the Bodies The nucleus pulposus becomes dehydrated and shrunken and the whole nucleus degenerates and narrows. Degenerated fibrocartilage, having none of the elasticity of a healthy disc, bulges beyond the rim. The bulge lifts periosteum from the bone, and periosteal new bone—an osteophyte—forms at the rim of the body

Protrusion of nucleus pulposus through a weakened annulus has the same effect. These protrusions are nearly always posterior and they are subsequently enclosed by osteophytes.

[1] McNew J C (1953) *Proc roy Soc Med.*, 46, 351

X ray films show osteophytes projecting from upper and lower margins of vertebral bodies, most often antero-laterally sometimes posteriorly They vary in size from slight lipping to large spurs of bone. Adjacent osteophytes may meet, but rarely fuse The corresponding disc space is narrowed [FIG 24]

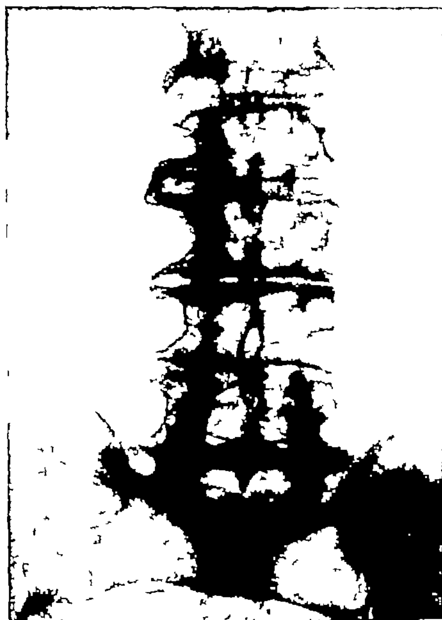


FIG 24

Lumbar spondylosis. Note beak-like osteophytes increased density of articular lamellae thinning of intervertebral discs.

Pain cannot arise from a degenerated disc for it is not provided with sensory nerves but there are three ways in which it can cause backache

1 from stretching of ligaments and periosteum by the bulging disc and from secondary chronic strain on more remote ligaments

2 from compression of nerve roots by posterior osteophytes

3 from secondary osteo-arthritic changes in the apophyseal joints

Spinal Osteo-arthritis At the level of the degenerated disc normal stresses and strains between the articular facets are inevitably altered this is a sufficient condition for the development of osteo-arthritic changes

Spinal osteo-arthritis is a name that should be restricted to osteo-arthritis of the apophyseal joints. Disc degeneration and osteophytosis of the bodies may exist without it and it may occur independently of disc degeneration.

Radiologically apophyseal osteo-arthritis will be detected in some 50 per cent. of instances of disc degeneration. An oblique view is usually best for its detection in the lumbar and dorsal regions. The characteristic appearances are narrowing of the joint space, which becomes indistinct marginal lipping and, the change most easily seen, a sclerosis of the subchondral bone.

Coccydynia

This is a persistent coccygeal pain In some few cases it is a sequel of injury—either a fracture or a sprain of the sacro-coccygeal ligament. It is prone to affect neurotic women, and here the cause may be psychogenic. It is also often associated with degeneration of the L.5-S.1 disc, and may be referred pain and tenderness from this lesion or the result of pressure on S.2 nerve root. Tender areas in the soft tissues on both sides of the coccyx may be found

Examination should include X ray films but no significance should be attached to asymmetry or unusual angulation of the coccyx

Spinal osteoporosis [p 37]

Post menopausal and senile osteoporosis are very common The complaint is of chronic backache, with a tendency to severe immobilizing paroxysms of pain in the lower dorsal or lumbar region, due to collapse of a vertebral body Severe root pain may follow collapse The range of movement in the spine is usually though not always, restricted

For differential diagnosis see pp 37–40

Paget's disease [p. 28]

In most cases there is complaint of chronic pain in the bones affected by this disease. It is a fairly common cause of low backache in patients over middle age.

PAIN FROM OTHER STRUCTURES

In many instances of low back pain we cannot demonstrate any disorder of the spine. For their explanation there are four possibilities

1 There is a spinal lesion though we cannot demonstrate it.
2. The pain is referred from a soft tissue lesion.
3 The pain is referred from visceral disease.
4 The pain is not referred pain

PAIN FROM UNDEMONSTRATED SPINAL LESION

Either we find no physical signs on examining the spine, or the signs we do find have no diagnostic power The possibility that even so there is a disc lesion cannot be denied with pain persisting after a typical disc protrusion there may be no diagnostic signs, but we infer from the history that some source of pain in the disc is still there.

There are some who assert that certain lumbar signs, such as a painful arc during some movement, indicate a minor disc protrusion or fragmentation But where their information comes from I do not know I suggest that for the present it be neglected

It has been suggested that the nucleus has a variable affinity for water and that if it becomes temporarily turgid confined as it is by the annulus and the vertebral bodies, an attack of lumbago is the result. This is an attractive hypothesis that could account for many puzzling attacks

PAIN REFERRED FROM A SOFT TISSUE LESION

Sprain

If a man falls heavily in a sitting position, he may sustain, in order of decreasing probability 1 a myofascial sprain 2. a sprain of sacro-iliac or intervertebral ligaments 3 an injury to a healthy intervertebral disc.

Sprains are most commonly of superficial structures, and therefore the ensuing pain is fairly sharply localized to the site of the injury It may last some 4 weeks or more the constancy in the site of pain and tenderness is noticeable and of diagnostic value.

Sprains of deeper structures are associated with the phenomena of referred pain pain is diffuse and very poorly localized it may be felt not only in the back, but in the hip and thigh, far from the site of injury In the painful area may be found a number of points tender to finger tip pressure.

Postural strain

Under this heading we include muscular and ligamentous strain from

faulty posture and general fatigue sacro-iliac strain following parturition chronic strain from spinal deformities and possibly some congenital abnormalities.

Faulty Posture A sagging posture, from increase in the normal curves of the spine, is seen much more frequently in the slender type of body build than in the average type. In most instances bad postural habits begin in childhood and persist throughout life but only in adult life do symptoms of ligamentous and muscular strain appear

Sacro-Iliac Strain. The sacro-iliac ligaments are normally of great strength, and that low backache can arise from strain is debatable. It is possible 1 from increased burden on a healthy ligament this would be imposed by rapid increase in weight or unaccustomed prolonged standing in a stooping position 2. from a normal burden on a ligament that has lost some of its tensile strength this occurs in the later months of pregnancy

Many diagnostic manoeuvres have been described for throwing a strain on the sacro-iliac ligaments without causing movement at the lumbo-sacral junction. One of the best is to test rotation of the trunk with the patient sitting, a unilateral pain on rotation being suggestive of a sacro-iliac lesion.

Spinal Deformities The idiopathic scoliosis beginning in childhood is as a rule painless until adult life, when heavier and more prolonged physical exertions bring the chronic pain of ligamentous and muscular strain

Spondylolisthesis. This is a fairly common developmental error in which the neural arch fails to fuse with the vertebral body The 5th lumbar vertebra is usually affected, sometimes the 4th, and very occasionally another It allows the vertebral column to slide forwards on the vertebra below the defective vertebra a condition called spondylolisthesis. The amount of forward slipping is very variable, but even in minor degree is probably a sufficient cause of low backache.

The patient is usually a young adult, male rather more often than female. A clue to the condition may often be had from inspection of the back, in a marked depression just above the 5th lumbar spine. If forward displacement is marked, there is a compensating lumbar lordosis and often a more vertical sacrum from backward rotation of the pelvis can be detected.

The diagnosis is confirmed radiologically all but minor degrees of displacement being clearly visible in lateral films.

Minor Congenital Defects. Such abnormalities in the lumbosacral region are quite commonly seen in X ray films. There may be obliquity of the plane of the apophyseal joints what should be a 5th lumbar

vertebra fused with the sacrum what should be a first sacral vertebra forming a 6th lumbar vertebra unilateral sacralization of the 5th lumbar vertebra and many other abnormalities. They are sometimes held responsible for low backache, on no very good evidence.

PAIN REFERRED FROM VISCERAL DISEASE

The phenomena of referred visceral and referred somatic pain appear to be identical

Lumbar pain

Pain in the upper lumbar region and loin a constant dull ache or attacks of sharp pain, is a common early symptom of *renal calculus* Red and white blood cells will probably be found in the urinary deposit. X ray films of the lumbar spine will show except with the rare calculus of pure uric acid, a shadow in the renal area in the A.P view lateral to the upper lumbar vertebrae, usually the 2nd in the lateral view superimposed on the vertebral body Renal colic occurs when a calculus (or blood clot) passes down the ureter excruciating pain shooting from the upper lumbar region, over the loin and down to the groin, comes in paroxysms and lasts perhaps several hours. An attack is commonly followed by haematuria.

Malignant growth of the kidney beginning usually after middle age, is accompanied by upper lumbar pain and pain in the loin but its prominent symptom is haematuria. Attacks of renal colic may occur from the passage of blood clots.

Chronic pyelonephritis is responsible for lumbar pain and marked general fatigue as well as frequency of micturition

Perinephric abscess in its early stages is accompanied by pain in the lumbar region and loin, the hip and the thigh. Later the increasing toxaemia, profuse sweats and high swinging pyrexia, with tenderness and bulging in the posterior renal angle, may suggest the probable diagnosis

Constant dull lower lumbar pain is associated with *chronic salpingitis* but so also are menorrhagia and dysmenorrhoea which give the indication for the necessary investigations.

Sacral pain

Sacral pain in women must suggest the possibility of *pelvic disease* but it is improbable that this will be the only or the most prominent symptom

Although sacral pain is associated with *carcinoma of the uterus*—late in carcinoma of the cervix, early in carcinoma of the body—irregular bleeding is the presenting symptom.

Pelvic pressure from *ovarian tumour* is rare and here sacral pain will be associated with difficulty of micturition and perhaps oedema of the legs.

Uterine fibroid rarely causes pain, but occasionally a dull backache is complained of even so the more prominent symptom is likely to be menorrhagia, the length of the period gradually increasing, and the long continued haemorrhage leading to chronic ill health from secondary anaemia.

Retroversion of the uterus is often symptomless. If it is a cause of sacral backache, as any cause of uterine congestion may be it will be associated also with menorrhagia, discharge, and dysmenorrhoea.

Prolapse of the uterus causes pain in the groins and sacral backache but at the same time there is frequency or difficulty of micturition and stress incontinence.

In reviewing the possibility of disease of the pelvic organs carcinoma of the prostate and of the rectum may have to be considered

Carcinoma of the prostate causes, first, difficulty in micturition, later frequency and precipitancy and sometimes chronic retention. It is accompanied by dull aching low back pain, and discomfort in the perineum.

Carcinoma of the rectum gives a dull aching rectal and sacral pain. Suggestive symptoms are wasting and diarrhoea in a middle-aged patient. He is likely to complain of several bowel actions during the day each action scanty and explosive. Haemorrhage from the bowel is common.

Pelvic spasm from any cause will give referred lumbosacral pain. *Spastic colon* is no exception. It is a characteristic manifestation of 'vagotonia'—the autonomic disturbance with excess of parasympathetic activity which is the common somatic manifestation of anxiety neurosis. Its clinical sign is a tender hard colon palpable in the left iliac fossa. Its prominent symptoms are an aching lower abdomen accompanied by lumbosacral pain. Low back pain is almost constant it varies in severity with that of the abdominal pain. If low abdominal pain is relieved by sedatives and antispasmodics, the lumbar pain is relieved at the same time. The condition is often associated with many other symptoms of autonomic disturbance.

PAIN WHICH IS NOT REFERRED

What remains to be said is still debatable. There is, however evidence that many instances of lumbago with its diffuse pain and focal tenderness, are not instances of referred pain.

Virus myalgia

Severe lumbar pain is a well-known feature of a number of acute virus infections. It is prominent at the onset of influenza, and variola, and

poliomyelitis It was characteristic of the 'trench fever of the First World War a virus infection transmitted by lice and now extinct.

Epidemic myalgia [p 142] is the only proved instance of a myositis caused by virus infection It seems highly probable however that epidemics of lumbago of short duration are quite common and that they are due to infection by a similar myotropic virus. In one recorded epidemic many cases were associated at the onset with a common cold.

Myalgia of anxiety neurosis and psychogenic pain

A diffuse aching in the lumbar region with localized zones of deep tenderness in the painful area, may be a part of that general rheumatic myalgia that is such a common complaint in anxiety neurosis [p. 4]. Diagnosis rests on the exclusion of other causes, on the peculiar features of this myalgia, and its associated symptoms, and on the mental symptoms of anxiety neurosis.

Psychogenic pain [p 5] may be prominent in any part of the spine. One form of hysterical pain in the back is the well-known railway spine or traumatic neurosis a complaint of persistent pain following a comparatively slight injury for which compensation is possible. Even some years after the injury the patient may still be complaining of agonizing pain and sleepless nights.

DORSAL PAIN

[See also Intercostal Pain, p. 139]

As in low back pain, we are guided by the probability that the source of pain is in the spinal column. Radiography includes the ribs If no disorder of the spine or ribs can be detected we then consider 1 a lesion of the spine though we cannot demonstrate it 2. pain referred from visceral disease 3 fibrositis, and psychogenic pain

The causes of dorsal pain are

SOMATIC

REFERRED FROM THE SPINAL COLUMN

Kyphosis

angular tuberculous metastatic carcinoma trauma etc.
smooth adolescent kyphosis senile kyphosis ankylosing spondylitis

Scoliosis

Disc pro

Spondyl

Rarefyin

FIBROSITIS [... e spine

rosi

VISCERAL

Pleural pain

Interscapular region carcinoma of the oesophagus mediastinal new growth aortic aneurysm

Inferior angle right scapula—T 9 level

gall bladder disease chronic cholecystitis biliary colic

T 12 level duodenal ulcer invading the pancreas carcinoma of the pancreas

PSYCHOGENIC

Hysteria

PAIN REFERRED FROM THE SPINAL COLUMN

Most of the spinal diseases discussed under low back pain may affect the dorsal spine.

Spondylosis

Spondylosis, i.e disc degeneration and osteophytosis, is common in the dorsal spine, especially in the lower part. Affecting the upper and middle portions in the elderly it is the cause of senile kyphosis [p 107] Osteo-arthritis of the apophyseal joints is also common in the dorsal spine, judging from autopsies but it is difficult to demonstrate radiologically

Rarefying diseases [pp 31–42] involve the dorsal as well as the lumbar spine.

Angular kyphosis

Any of the spinal diseases resulting in an angular kyphosis may cause a diffuse pain in the dorsal region with local tenderness, or may be a cause of intercostal neuralgia [p 140]

Angular kyphosis results from collapse of a vertebral body or several adjacent bodies The commoner causes are tuberculous osteitis [p. 91 & FIG 25] and carcinoma [p 93]

Falls from a height, landing on the feet or sitting, usually cause a *simple compression fracture* of several vertebral bodies. In X ray films one body appears markedly wedged, its anterior part being crushed vertically those immediately above and below are less severely compressed.

Kümmell's rarefying osteitis is a delayed wedging of a vertebral body after comparatively slight injury It probably follows an unsuspected crack fracture of the body Perhaps some months after apparent recovery localized aching pain is complained of Serial X ray films show a slowly developing rarefaction and wedging of one vertebral body



FIG. 25
Tuberculosis of the spine. Several vertebral bodies have collapsed, causing an angular kyphosis.

at this level It may be confused with tuberculosis, or a secondary deposit of carcinoma.

There are several rare causes of wedging of a single vertebral body *Syphilitic osteitis* may closely resemble Pott s disease clinically and radiologically so closely that diagnosis usually rests on other evidence of syphilis, including a positive Wassermann reaction. It is a late tertiary manifestation

Tabetic Charcot spine is a deformity of the spine due to bone destruction and compression of one or two vertebral bodies usually in the lower dorsal or lumbar region X ray changes are very marked bone destruction and considerable deposition of new bone There is local discomfort and perhaps root pain but no local tenderness or spasm Clinical evidence of tabes will be found [p 165]

Collapse may occur in *lymphadenoma* from deposits of lymphadenomatous tissue in the vertebral body the appearance radiologically resembling that of metastatic carcinoma. A similar change may occur in *leukaemia* and in *myelomatosis* beginning as a solitary tumour

Staphylococcal osteomyelitis is another rarity liable to confusion with tuberculosis [p 93]

There is a rare osteochondritis of the spine called *Calré's disease* It occurs in young children and closely mimics Pott s disease There is a progressive collapse of a single vertebral body till it becomes a flattened disc, with a consequent angular kyphos, and pain. It usually occurs in the dorso-lumbar region

Smooth kyphosis

The more important causes of a diffuse dorsal kyphosis are adolescent kyphosis, senile kyphosis, and ankylosing spondylitis the first two are due to changes in the intervertebral discs

Adolescent Kyphosis (*Scheuermann s Disease*) This affects mainly the mid- and lower thoracic spine. It is often discovered in adolescent boys and girls during a routine examination, before it has begun to cause aching. The kyphosis tends to increase up to the age of 21 and aching is usually complained of when boys begin to do heavier manual work.

The essential change which brings this deformity appears to be a series of Schmorl s nodes—nuclear prolapse into the adjacent vertebral bodies. The consequently narrowed discs lose elasticity and growth of the vertebral bodies is affected. In the anterior part especially is growth defective the epiphyseal rings are fragmented, and the bodies become wedge shaped.

Thus in a well established case of adolescent kyphosis X ray films of the dorsal spine show 1 a series of Schmorl s nodes 2. narrowing of the disc spaces 3 wedging of the vertebral bodies 4 fragmentation of the epiphyseal rings.

Senile Kyphosis This is an increase in the normal curve in the upper and mid-dorsal zones, fairly common in elderly people. Degenerative changes in the discs, especially in the anterior parts, allows forward tilting of the vertebrae, which may become slightly wedge-shaped Here and there two adjacent vertebral bodies may be seen, in lateral

radiographs, fused anteriorly Often bone density is normal, the deformity being due to disc degeneration and not to senile osteoporosis.

The condition is sometimes accompanied by severe dorsal aching.

Ankylosing Spondylitis [p 87] When the disease has affected the whole spine, some patients maintain a fairly good erect posture but more often a well-marked kyphosis in the upper dorsal spine has developed This deformity may be so severe as to compel the sufferer to look vertically downwards when he stands. The lumbar and lower half of the dorsal spine are almost in a straight line Pain in the dorsal region of the back in this late stage varies a good deal but there is usually some continual aching, and it may be severe Not infrequently it causes intercostal neuralgia [p 140]

Scoliosis

Postural Scoliosis This is a long gentle lateral curve, with slight bulging of the ribs on the side of the convexity When the spine is fully flexed the deformity disappears. This is one of the postural defects to which adolescents are prone It is usually temporary disappearing in a few years without treatment. Postural scoliosis rarely causes any discomfort

Such a lateral curve is also caused by shortening of one leg and consequent tilting of the pelvis here minor changes may in time occur in the shape of the vertebral bodies, but the curve can always be partially corrected

Structural Scoliosis. This is a more localized deformity with secondary curves above and below the primary one The ribs bulge prominently on the side of the convexity and the deformity does not disappear with flexion of the spine The common type is the idiopathic scoliosis beginning in childhood As a rule it is painless until adult life, when pain from chronic ligamentous and muscular strain or sometimes root pain, is apt to appear

PAIN REFERRED FROM VISCERAL DISEASE

Some of the visceral causes present no difficulty other symptoms being far more prominent and constant than pain in the back. But the only way to be reasonably sure of avoiding mistakes is 1 to note the presence of chest or abdominal symptoms, and to make examination of the chest and abdomen a routine procedure 2. In all cases of severe interscapular pain and in all doubtful cases, to get X ray films of the chest. It should be remembered that both pulmonary tuberculosis and bronchial carcinoma may be accompanied by mild and ill-defined pain

in the chest wall and that many people with persistent pain like this are anxious about the possibility of lung disease

Pleural pain

Pleural pain is felt over the area of pleura involved except when this is the dome of the diaphragm when it is referred to the C.4 segment in the cervico-scapular region It may be a symptom of several chest diseases.

Tuberculous Pleurisy This condition presents as pain most commonly in the lower axillary area, and near the inferior angle of the scapula. Pyrexia, rapid shallow breathing, diminished expansion of the chest and a friction rub are the data establishing the diagnosis The picture, however may be less definite than this and if pain persists in a young adult the chest may have to be examined repeatedly Residual pain from a pleurisy long since abated is fairly common Pain in the chest wall that not uncommonly accompanies pulmonary tuberculosis is probably of pleural origin.

Pulmonary Infarction Pleurisy accounts for the pain of pulmonary infarction arising for example in heart disease or thrombophlebitis.

Bronchial Carcinoma. Pleurisy also accounts for the chest pain in some instances of bronchial carcinoma in which a bronchus is blocked. In other instances of carcinoma the growth causes severe local pain by invading the ribs.

Chronic Bronchitis Episodes of aspiration pneumonia and pleurisy may occur in this condition

Interscapular pain

Carcinoma of Oesophagus. The salient feature of early carcinoma of the oesophagus is more and more frequently recurring difficulty in swallowing. Only occasionally is there pain this is usually felt behind the sternum, and is only occasionally interscapular

Mediastinal New Growth Severe interscapular pain may be caused by erosion of vertebrae by a mediastinal new growth but symptoms of pressure on the great vessels, the oesophagus and trachea are likely to be equally prominent.

Aneurysm of the Descending Aorta. This condition is rare Its most prominent symptom is extremely severe interscapular and girdle pain from erosion of vertebrae.

Pain at the inferior angle of the right scapula

Chronic Cholecystitis. This disease is associated with pain in the right upper abdominal quadrant, radiating through to the back just below the inferior angle of the right scapula. Sometimes it is felt most in the

back A history of mild attacks of biliary colic, a complaint of epigastric fullness and nausea after meals, and on examination tenderness in the right upper quadrant, should lead to the right investigations. The severe, even agonizing pain of *biliary colic* is felt in the epigastrium, or right hypochondrium, and radiates through to the back at this level.

Pain at the level of T.12

A chronic *duodenal ulcer* invading the pancreas will be a late event in a disease long since diagnosed

Carcinoma of the pancreas is characterized by a progressive jaundice, with anaemia, nausea and loss of weight In some cases there is deep epigastric pain radiating through to the back.

PAIN IN THE ELBOW

Although arthritis is a possibility the cause of a painful elbow is usually outside the joint and it may be in the neck and shoulder region.

LOCAL CAUSES

lateral epicondylitis— tennis elbow
olecranon bursitis
arthritis rheumatoid tuberculous osteo-arthritis
loose bodies osteochondritis dissecans

REFERRED PAIN

from a lesion in structures supplied by C.5 T.2

PAIN FROM LOCAL CAUSES

Tennis elbow

This is the commonest of the local causes of a painful elbow There is no more than some aching at rest but sharp pain is evoked by a strong grip

There is no local swelling, and passive movements at the elbow are of full range. Palpation reveals deep tenderness over the lateral epicondyle of the humerus, and perhaps also over the head of the radius. Every case shows the following phenomenon ask the patient to hold out the hand with straight fingers, and to resist your attempt to flex his fingers as he brings into action his extensor muscles, your attempt to flex his middle finger at the metacarpo-phalangeal joint will produce pain at his elbow

For the condition is, it is generally thought, a chronic sprain of the origin of the extensor muscles from the lateral epicondyle of the humerus. It would be better called lateral epicondylitis A grip involves contraction of the extensors of the wrist and fingers, as fixator muscles

otherwise flexion of the fingers would also produce flexion at the wrist Every grip is therefore apt to be severely painful at the elbow

Arthritis of the elbow Joint

The joint is swollen, all movements are restricted in some degree and muscular wasting especially in the upper arm is marked

Tuberculous Arthritis In children and adolescents the commonest arthritis of the elbow is tuberculous arthritis. The joint gradually becomes stiff swollen and tender pain is not as a rule severe, though some patients experience night starts Infection often begins in the olecranon little more than local tenderness will be found at this stage. But the joint is soon involved and the characteristic doughy white swelling appears—compounded of tuberculous material distending the synovial cavity and infiltration of the synovial membrane and peri-articular tissues. The joint swelling is all the more obvious because of the rapid wasting of arm and forearm muscles. Later a cold abscess may form by the olecranon, breaking the skin and leaving a sinus. The X-ray appearances are erosion of joint surfaces marked osteoporosis, destruction of olecranon and coronoid processes.

Rheumatoid Arthritis. A tuberculous elbow is a monarticular affection the other joints are healthy It must be very exceptional for rheumatoid arthritis [p 190]—the commonest arthritis of the elbow in adults—to begin in the elbow and still more rare for it to remain confined to this joint very long. Such cases would present some difficulty in diagnosis. Commonly however the elbow is affected by rheumatoid disease after fingers, wrists, and other joints, and when the diagnosis has been made

Osteo-arthritis Osteo-arthritis of the elbow is uncommon often there is a history of injury to the joint or of such repeated minor traumata as those to which boxers, or pneumatic drill workers, are subject. Pain may be felt only on certain movements at the joint but it may be continual, and is sometimes quite severe. There may be complaint of locking, from the presence of a loose body

Unless osteophytosis is extensive, swelling is not so marked as in a true arthritis of the joint. As a rule no capsular thickening can be made out, and there is no peri-articular infiltration The swelling is largely bony with perhaps slight synovial effusion It is made more prominent by muscular wasting in the arm and forearm Occasionally osteophytes at the medial side of the humero-ulnar joint cause ulnar paralysis from ulnar nerve compression. The X ray appearances of the joint are quite characteristic and no difficulty in diagnosis arises when films are obtained as a routine in all painful elbows whose movement is restricted [FIG 26]

Osteochondritis Dissecans This condition affects chiefly the knee and elbow the incidence is said to be 80 per cent in the knee and 10 per cent in the elbow A disc of the articular lamella is cut off from the rest of the articular bone, either of the capitulum or of the head of the radius, by a thin arc of granulation tissue and with its covering cartilage it eventually separates to form a loose body in the joint, leaving behind it a saucer-shaped depression.

The disorder is one of adolescence and young adult life, and therefore must be distinguished from early tuberculosis. The features are very



FIG 26
Osteo-arthritis of the elbow

different. Before the fragment has separated there may be moderate pain and restricted movement, slight local tenderness and slight swelling from synovial effusion. But there may be no symptoms at all until the presence of a loose body in the joint makes itself known by an episode of locking. X ray films show the separating fragment, or the depression it previously occupied.

Loose Bodies Loose bodies in the elbow are not uncommon The source may be osteochondritis dissecans, or a detached osteophyte in osteo-arthritis and synovial chondromatosis may be responsible for multiple loose bodies.

The only symptoms may be increasing pain and stiffness. The suggestive clinical feature is locking—but this only occurs when the fragment is nipped, during joint movement, either between the olecranon posteriorly or the coronoid process anteriorly and the humerus. The

fragment may not be in a position to interfere with movement. It is easy to overlook a small, loose body in X ray films.

REFERRED PAIN

Almost any cause of pain in the neck and shoulder may cause referred pain in the arm But as a rule pain is not *felt equally in the whole extent* of the somatome. Maximum pain is felt in one or more localized zones often in the region of the elbow

Referred pain at the elbow will almost always be accompanied by pain in the cervico-scapular region. Because of referred tenderness it may easily be confused with tennis elbow But tenderness is less localized, and the finger test is negative.

PAIN IN THE FOOT AND ANKLE

DIFFUSE PAIN

Of disorders tending to cause diffuse pain rather than pain localized to one part of the foot, the more important are

ARTERIAL DISEASE
- *sudden arterial occlusion*
- *intermittent claudication*
- *rest pain from ischaemia*
- *erythromelalgia*

RHEUMATOID ARTHRITIS

ARTERIAL DISEASE

The patient's account of the pain which is due to *sudden arterial occlusion* is almost diagnostic. It is usually of sudden onset, continuous and cramp-like in the foot and leg and it is accompanied and sometimes preceded, by numbness and coldness. The skin of the foot and lower part of the leg is blanched and later mottled with cyanotic patches The simpler tests for arterial deficiency should be carried out [p 143]

Occlusive arterial disease

Either arteriosclerosis obliterans, or thrombo-angiitis obliterans, may be responsible for

1 Intermittent claudication [p 142] Though pain is most common in one calf it is sometimes felt in one foot.

2 Rest pain from ischaemia. This is felt only in advanced arterial

disease, as a dull continuous aching pain in the foot and lower part of the leg, common at night and badly disturbing sleep

Erythromelalgia

This condition is erythralgia [p 45] in chronic form affecting the feet. It occurs in the decline of life the well known senile burning paraesthesia of the feet The complaint is of severe burning pain, induced by moderate warmth such as warm water or the warmth of a bed, or by the friction inseparable from walking Walking for some patients is too painful to be attempted Relief is continually sought by



FIG. 27
Rheumatoid arthritis of the ankle.

cooling the feet. The skin is red and tender and may feel colder or hotter than normal.

Some instances are associated with occlusive arterial disease but in many there is no evidence of this.

RHEUMATOID ARTHRITIS

When rheumatoid arthritis begins, as it may in the joints of the foot, diagnosis in the early stage is always difficult. Pain is often felt only in the metatarso-phalangeal region but if the tarsal joints are also affected it will be felt in the whole foot.

A fully developed arthritis, with its slight general soft tissue swelling,

the painfully restricted passive movements of toe and tarsal joints radiological porosis of the bones of the foot and perhaps one or two small joint erosions, is easy to recognize [110 27] Before this a tentative diagnosis rests on the exclusion of other causes of painful feet and the recognition of the early rheumatoid clinical picture [p 190] Rheumatoid arthritis does not long remain confined to the joints of the foot

LOCAL PAIN

PAIN IN THE METATARSO PHALANGEAL REGION

The causes may be grouped as follows

TOES

Hallux valgus hallux rigidus osteo-arthritis of the sesamoids claw foot unsuitable shoes Morton s metatarsalgia minor deformities and secondary corns ingrowing toe nail subungual exostosis

METATARSALS

March fracture osteochondritis of the metatarsal head osteoid osteoma

METATARSO-PHALANGEAL JOINTS

Rheumatoid arthritis gout

SOFT TISSUES

Plantar warts arterial disease Raynaud's disease with trophic changes incipient gangrene

Metatarsalgia is usually caused by throwing most of the weight, when standing and walking on the outer part of the foot painful disorders of the big toe will have this effect. Normally most of the weight is taken by the more massive bones of the arch on the inner side of the foot

Most of the conditions responsible present no diagnostic difficulty A cause sometimes overlooked is unsuitable shoes The chief offender is the type of shoe that allows the toes to be thrust forwards against the toe-cap at every step the high-heeled shoe with no heel platform any shoe that exerts no counter thrust at the ankle or tarsus to the natural forward slip of the foot in the shoe when walking. A boot most effectively avoids this antero-posterior compression of the toes

Pain from postural defects when moderate is in the region of the metatarsal heads when severe it may affect the whole foot and even the calf On examination, toes will be found clawed the metatarsal heads prominent, and tender to pressure on the plantar surface There may be some general swelling of the metatarsal region the plantar surface of which is concave from side to side.

Morton's metatarsalgia

This is an uncommon condition, due to a local enlargement of a plantar digital nerve. The patient suffers attacks of very severe pain in the cleft between the 3rd and 4th toes and shooting into the toes, coming on for no apparent reason. The foot is pain free in the intervals. The characteristic sign is pain on upward and backward pressure at the base of the 3rd and 4th toes Pain in the cleft is also felt on lateral compression of the anterior part of the foot. It is due to the presence of



(*a*)



(*b*)

FIG. 28

March fracture

(by kind permission of Dr G A S Lloyd).

(*a*) Soon after onset of pain and tenderness over 2nd metatarsal no change visible.

(*b*) Two weeks later callus at the site of fracture.

a plantar digital neuroma—a fibrous fusiform swelling of the plantar digital nerve as it approaches the interdigital cleft between the 3rd and 4th toes. The condition is completely relieved by its resection The swelling is most probably the result of ischaemia the digital artery is found degenerated The same condition may occur though less commonly in the 2nd cleft

Affections of the metatarsals

When a painful foot is found on examination to show a point of acute tenderness over the shaft of a metatarsal X ray films should be obtained Only so can a march fracture be detected It affects any of the outer 4 metatarsals, usually the 2nd or 3rd There is complaint, after a long walk, of pain or an indefinite aching in the metatarsal region and on examination there will be found oedema of the dorsum, and localized tenderness over the shaft of one of the metatarsals. The onset is likely to be insidious, beginning with an indefinite ache and increasing to severe pain. An X ray film early may show no change, but after 2 or 3 weeks a fusiform deposit of new periosteal bone will be seen around a part of the shaft of the tender metatarsal A fracture line may or may not be seen running across it [FIG 28] The nature of the condition is a little obscure.

Osteoid Osteoma [p. 26]. This tumour has been described affecting metatarsals and phalanges, and is a possible cause of pain and local tenderness. Seventy five per cent. of recorded cases have been in patients between the ages of 10 and 25 years. It is suspected by the radiological appearances, and proved only by biopsy

Osteochondritis of a Metatarsal Head This is an uncommon condition seen only in adolescents The complaint is of aching in the fore part of the foot The condition affects the head of the 2nd or 3rd metatarsal, which is enlarged and tender X ray films show it to be broad and fragmented [FIG 29]

Metatarso-phalangeal joints

Rheumatoid Arthritis This disease may begin in these joints [p 191]

Gout The classical attack of acute gout in the big toe should present no diagnostic difficulty It usually begins in the night, with severe pain in the ball of the big toe. Pain abates a little in the morning, but the toe will be found swollen, red and shiny The skin is bright red for the first 24 hours it then develops a violet hue, and oedema increases. For some 4 days severe pain continues, waxing and waning slightly It then gradually lessens, and after another week or so nothing remains but some local discomfort redness and swelling disappear stiffness passes

off These attacks tend to recur perhaps once or twice a year in more fortunate patients at intervals of some years.

If there is any doubt from the appearances of the joint, remember that

1 men are attacked far oftener than women
2 the first attack is not often at an earlier age than 35
3 there is often a family history of gout
4 there may be tophi in the rim of the ear though this is unlikely at the time of the first acute attack
5 the serum uric acid is likely to be in the region of 6 mg. per 100 ml., or above.



FIG. 79

Osteochondritis of a metatarsal head (by kind permission of Dr Cecil Bull).

Soft tissues

The possibility of metatarsal pain being due to *plantar warts* should not be overlooked. They may form anywhere on the sole but are common under the ball of the foot. They project but little if at all from the skin, and close inspection is needed They are exceedingly tender

Constriction of the small arteries and arterioles does not cause much pain but pain in the toes and fore part of the foot may be quite bad in severe forms of Raynaud's disease especially when associated with trophic changes

PAIN IN THE TARSAL REGION

The chief causes are

PES VALGUS

ARTHRITIS OF THE TARSAL JOINTS
rheumatoid gonococcal due to Reiter s disease due to gout tuberculous

OSTEO-ARTHRITIS OF THE TARSAL JOINTS

KÖHLER S DISEASE (osteochondritis of the navicular)

Pes valgus

This deformity is not a sufficient diagnosis We want to know

1 *If there is another cause of pain* Quite often a valgus foot is painless—but such feet are fully mobile. A foot painful because of the valgus deformity shows some loss of mobility If therefore a valgus foot is fully mobile it is well to consider other possible causes of pain.

2. *The cause of the valgus deformity* The causes are reviewed on p 55 Particularly do we want to sort out from the rest the spastic flat foot of adolescents, and the pes valgus of rheumatoid arthritis.

Pain from the abnormal posture comes from chronic ligamentous strain. It is at first a dull aching felt as a rule on the medial side of the foot near the first prominence below and in front of the medial malleolus, which is the tubercle of the navicular Sometimes pain is more marked under the heel or even the sole of the foot generally Rest brings relief

Arthritis of the tarsal joints

A severe and immobile valgus deformity is likely to be the result of *rheumatoid arthritis* affecting the tarsal joints.

An attack of *acute gout* does not necessarily fall on the big toe It may affect the tarsal joints.

Gonococcal Arthritis This may closely mimic gout. But it does not show the daily remission of the classical gouty paroxysm nor does it tend to abate after a few days under adequate doses of colchicine nor does it subside completely in one or two weeks as do most attacks of gout. It may cause a *considerable tender swelling* in the tarsal region. Remember that acute arthritis following gonorrhoea may be Reiter s disease [p 207].

Tuberculosis. Tuberculosis of ankle and tarsal joints is uncommon Essentially a disease of children it may occur in young adults and in the elderly The child develops a limp One foot is painful and found to be swollen, with a doughy swelling in the region of the ankle, or outer or

inner side of the tarsus or synovial distension of the ankle joint palpable on each side of the tendo Achillis, and later under the tendons anteriorly There is slight plantar flexion deformity and markedly restricted movement at ankle and tarsal joints. This clinical picture in children and adolescents means tuberculosis. Confirmation may be obtained by X ray films, which show evidence of bone destruction in one of the tarsal bones.

Osteo-arthritis Osteo-arthritis of the tarsal joints commonly develops in old valgus foot deformities Osteo-arthritis of the sub-taloid joint is a common sequel of injury notably of crush fractures of the calcaneus. X ray appearances are characteristic.

Osteochondritis of the Navicular (Köhler s Disease). This is seen only in children more often boys, the maximum incidence being 4-6 years. The child complains of aching pain on the inner side of the foot, and develops a limp. There may or may not be slight swelling and tenderness over the navicular X ray films, by which alone the diagnosis can be made, show the navicular to be flattened antero-posteriorly and increased in density

Pain abates in 2 or 3 months, especially if weight-bearing is reduced to a minimum the bone eventually acquires a normal structure.

The condition is one of a group of small bone and epiphyseal disorders named osteochondritis, although nothing is known of its pathology Köhler also described an osteochondritis of the metatarsal head [p 117]

PAIN IN THE ANKLE AND HEEL

The more important causes are

ANKLE

Arthritis of the ankle or talo-calcanean joint
rheumatoid tuberculous gonococcal due to Reiter s disease
Osteo-arthritis
Injuries sprain rupture of the external lateral ligament
Tenosynovitis
Synovioma

HEEL

Plantar fasciitis
Achilles bursitis
Achilles tendinitis
Apophysitis of the calcaneus
Plantar warts

PAIN IN THE ANKLE

A painful ankle from *arthritis of the talo-tibial or sub-taloid joint* presents no diagnostic difficulty other than that of distinguishing the several possible types of it Examination of the ankle region will show 1 some degree of plantar flexion deformity 2 restricted movement at the ankle joint or restricted inversion and eversion of the foot 3 swelling on each side of the tendo Achillis or below and in front of the malleoli 4 characteristic changes in X ray films, which should be obtained in all cases where movement is restricted

In children and young people *tuberculosis* is the commonest cause in adults *rheumatoid arthritis*

Sprained ankle

This follows acute overstretching of the middle or anterior fasciculus of the external lateral ligament In the majority there is probably no macroscopic tearing of the ligament, but only injury to sensory nerve endings but tears of any degree may occur to complete rupture of the ligament. The cause is a violent wrench of the foot inwards the immediate result a sharp pain on the outer side of the joint, and swelling below and in front of the external malleolus If the sprain is not intelligently treated the ankle may continue to feel painful insecure and stiff for many weeks or even months—a condition called chronic sprain, in which there may be found oedema around the malleoli or on the dorsum of the foot and restricted movements

The symptoms and signs of chronic sprain may be due to complete rupture of the external lateral ligament, with consequent continual giving way of the ankle—a recurrent subluxation It is detected by antero-posterior radiographs, with the foot held in full inversion.

Tenosynovitis

Tenosynovitis is a common disability caused as a rule by strenuous exercise after months of inactivity It can be recognized by a slight tenderness along the course of the tendons. It may affect the tendon sheaths on the dorsal aspect of the ankle it may affect the sheaths of the tibialis posterior and flexor tendons behind the medial malleolus—a common cause of disabling pain in ballet dancers.

Synovioma [p 128]

This tumour of synovial membrane may grow from tendon sheaths, and may be found in any of those at the ankle, causing aching pain and restricted movement.

PAIN IN THE HEEL

If pain is sharply localized under the heel the most probable cause is chronic strain of the attachment of the plantar fascia to the calcaneus —*plantar fasciitis* Plantar fasciitis, with periostitis of the os calcis and a painful calcanean spur is commonly found in *Reiter's disease* whether there is a true gonococcal plantar fasciitis is now doubtful.

Pain under the heel may be due to *plantar warts* which are acutely tender to touch. Although somewhat buried under the horny layer they are not as a rule difficult to recognize.

Pain behind the heel is a common complaint, that may arise from one of several conditions

1 A chronic inflammation of subcutaneous tissues from pressure of the hard heel of a too tightly fitting shoe.

2. A *bursitis* affecting either the superficial bursa between the Achilles tendon and the skin or the deep bursa between the tendon and the posterior surface of the calcaneus. The cause may be traumatic, or gout.

3 A *tendinitis* of the tendo Achillis, akin to tenosynovitis in being usually due to the trauma of overuse.

4 In children, especially in boys from 8 to 14 years old, the possibility of *apophysitis of the calcaneus* must be considered This is an instance of the group of disorders called *osteochondritis*, affecting here the epiphysis on the posterior aspect of the calcaneus. The back of the heel will be found swollen and tender X ray films show fragmentation and increased density of the epiphysis

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PAIN IN THE HAND

Complaint of a painful hand is very common It can be a source of considerable alarm in pianists, violinists, surgeons and others whose livelihood depends on manual dexterity And so advice is sought for many minor ailments and the proof that they are minor is often troublesome

The usual cause is some local lesion, readily detected by simple inspection, and palpation testing movements, and a radiograph When no local cause can be found it is possible that we are dealing with some nervous or vascular cause of pain in the upper limb felt, as is sometimes the case, chiefly in the hand In the usual forms of brachial neuralgia

it is unlikely that pain would be confined to the hand but painful paraesthesia may be

The chief causes are

LOCAL LESIONS

Skin local septic and ulcerative conditions Raynaud s disease sclerodactyly

Tendon sheaths tenosynovitis Quervain s disease ganglion synovioma

Joints osteo-arthritis arthritis—rheumatoid gout, gonococcal Reiter s disease tuberculous, hypertrophic osteo-arthropathy

Bone tubercle enchondroma solitary cyst osteoid osteoma Kienböck s disease Madelung s deformity

NERVOUS AND VASCULAR CAUSES OF UPPER LIMB PAIN

Reflex sympathetic dystrophy [partial shoulder hand syndrome p 83]

Thrombo-angiitis obliterans

PAINFUL PARAESTHESIA

Brachial neuralgia [p 74]

Acroparaesthesia [p 186]

There are some diagnostic problems that arise repeatedly among them 1 the recognition of early rheumatoid arthritis 2. the differential diagnosis of tenosynovitis 3 the diagnosis of a painful hand showing trophic changes.

THE RHEUMATOID HAND—DIFFERENTIAL DIAGNOSIS

The diagnosis of rheumatoid arthritis is considered in the article on polyarthritis [p 189] The rheumatoid hand, though very distinctive [FIG 30], may on occasion be confused with the following

Deformities of the Hand. See pp 56-8

Osteo-arthritis

Common in women over 40 the incidence increases with advancing age. It may affect any joint, but most often the terminal interphalangeal joints, and the carpo-metacarpal joint of the thumb The first indication is the appearance of little nodular swellings over the terminal joints, called Heberden s nodes They are soft tissue swellings over small osteophytes usually painless, they are often accompanied by stiffness and discomfort. When the morbid changes become more pronounced there may be quite troublesome pain in the fingers with marked swelling and increasing stiffness of the affected joints, and it may be some degree of flexion deformity [FIG 31]

The distinction from rheumatoid arthritis is not as a rule difficult. The joint swelling is irregular, hard and nodular in contrast to the softer capsular and peri-articular swelling of rheumatoid arthritis. The skin of the hand is normal and there is none of the cold clammy feel associated with rheumatoid arthritis nor any marked muscular



FIG. 30
Severe deformity of the hands in advanced rheumatoid arthritis.



FIG. 31
Heberden's nodes, and osteo-arthritis of the first carpo-metacarpal joint.

wasting nor any ulnar deviation of the fingers. Moreover rheumatoid arthritis hardly ever affects the terminal interphalangeal joints. The X ray appearances will confirm the clinical diagnosis. The E.S.R. is almost always normal in osteo-arthritis.

Garrod's pads

These are circumscribed thickenings of the soft tissues on the dorsal aspect of quite normal, though perhaps unduly prominent, proximal interphalangeal joints. They are not painful or tender there are no clinical signs in the joints, and X ray films are normal Their origin is unknown, and except for being a little unsightly they are of no importance



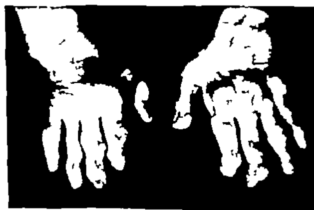


FIG. 32
Advanced gout with tophi
(by kind permission of Dr O Savage)

Chronic polyarticular gout

This disease in most instances affects the hands, as well perhaps as other joints. The hand may not be unlike that of rheumatoid arthritis but the joint swelling is harder and more nodular both proximal and

terminal interphalangeal joints are affected, there is little muscular wasting and no ulnar deviation. The only pathognomonic sign is the peri articular tophus on the dorsum of the finger joints but there may be none in the early stages. The pearly white colour of the sodium biurate of which it is composed can often be made out through its covering of thin skin. It is harder than the rubbery rheumatoid nodule, which may also form on the fingers and it is not necessarily over the joint line as is a Heberden node. Tophi vary in size from a pin s head to a hen s egg skin often ulcerates over the larger concretions [FIG 32] Some of the contents of these ulcerated tophi may be teased out with a needle and subjected to microscopic examination and the murexide test. Typical needle-shaped crystals of sodium biurate may be demonstrated the material mixed with 5 drops of dilute nitric acid is evaporated to dryness, cooled, and a few drops of ammonia added a purple colour will form if the material is sodium biurate.

When tophi cannot be recognized the most reliable sign is hyper uricaemia the plasma uric acid is 6 mg. per 100 ml., or more.

X ray films of the hands show small punched-out erosions near the articular rim in the phalanges or metacarpals or carpal bones. There will be some loss of cartilage and osteophytosis in advanced cases. Punched-out erosions do not occur in primary osteo-arthritis but they are often seen in rheumatoid arthritis, and are therefore not pathognomonic of gout. But some degree of osteoporosis is invariable in rheumatoid arthritis, and does not occur in gout.

Chronic polyarthritis associated with gonorrhoea

This condition may affect the small joints of the hand, the carpus and wrist. But most instances, if not all, are Reiter s disease, and not due to gonococcal infection of the joints [p 207].

Tuberculous polyarthritis

This is a rarity a form resembling rheumatoid arthritis has been described but its existence is doubtful. Rheumatoid arthritis may affect the carpal and wrist joints only Tuberculous infection of these joints occurs in children and young people but it is rare, and whereas the rheumatoid condition is nearly always bilateral, tuberculous disease affects only one wrist.

Hypertrophic osteo-arthropathy

Hypertrophic osteo-arthropathy [p 205] is often for a time treated as rheumatoid arthritis It is important to make the correct diagnosis, because now and then it is the first indication of a bronchial carcinoma.

AFFECTIONS OF TENDON SHEATHS

Polytenosynovitis

This condition may be a manifestation of several of the rheumatic diseases It commonly accompanies rheumatoid arthritis and sometimes in the early stages the impact of the disease seems to be more on tendon sheaths than on joints. It affects the hands and feet usually symmetrically The tendon sheaths, especially those around the wrist and the flexor sheaths of the fingers become swollen from an extensive proliferation of synovial villi and a sterile serous exudate The tendons in the palm may become thickened and nodular A nodule may interfere with active flexion, and may lead to a trigger finger

It may be seen also in *rheumatic fever gonococcal infections* and *gout*

The common *traumatic tenosynovitis* comes from overuse of the hand or more obvious strain It affects the hand chiefly at the wrist causing aching pain, which when severe spreads to forearm and hand Usually it is accompanied by no physical sign other than a fine creaking on moving the fingers.

Tuberculous tenosynovitis

This disease causes a painless chronic effusion, usually in the flexor tendon sheaths. It may be confined to one finger or it may spread to the flexor sheath in the palm and wrist, giving a swelling in the palm and above the wrist. Less commonly the extensor sheaths are affected.

Tenosynovitis stenosans

This is a localized thickening of a flexor tendon sheath, and consequent narrowing of its lumen, either of a finger—usually index or middle—or a thumb at the level of the metacarpo-phalangeal joint. There is also a localized nodular thickening of the tendon just distal to this narrow part of the sheath In flexing and extending the digit the nodule in the tendon must pass through the constricted sheath it does so with a snap—the so-called trigger finger or 'trigger thumb The power of flexion being stronger than that of extension active flexion is often possible but active extension not the flexed digit must be snapped back into extension with the other hand

Quervain s syndrome

This is a localized tenovaginitis affecting the common sheath of the abductor pollicis longus and extensor pollicis brevis. There is complaint of pain at the base of the thumb spreading to the forearm mainly when using the hand There is tenderness over the lateral side of the lower end of the radius, and some degree of swelling of the tendon sheath is usually seen here These two signs distinguish it from osteo-arthritis of

the carpo-metacarpal region of the thumb the commonest cause of pain at the base of the thumb

Ganglion

These well known cysts which appear at the wrist and hand or ankle and foot, filled with grey jelly are attached by a fibrous band to a tendon sheath or joint capsule. There is rarely any direct communication. It is uncertain whether they are herniations of a tendon sheath or synovial cavity or benign cystic neoplasms. They give little trouble, other than an unsightly swelling and some aching

Synovioma

There is a benign synovioma of tendon sheaths, most often found as a firm localized encapsulated tumour growing outwards from the flexor tendon sheath of a finger or thumb usually in young adults. They are very slow growing the only symptom is some restriction of movement.

Malignant synovioma (synovial sarcoma) is a highly malignant tumour also found most often in young adults. This usually arises from a joint or bursa common sites being knee ankle, foot, elbow wrist, hand 50 per cent. arise from the knee joint. But there is a diffuse malignant synovioma of tendon sheaths They are slow growing at first, but eventually cause considerable swelling liable to be mistaken for a lipoma. The tumour causes some pain and aching in the affected limb. Metastases occur in the lungs

PAINFUL HANDS WITH VASOMOTOR AND TROPHIC CHANGES

The hand in rheumatoid arthritis usually shows vasomotor and trophic changes in moderate degree the skin is cold and clammy slightly cyanosed shiny and thin.

The distinguishing features of Raynaud s disease and of sclerodactyly have been described elsewhere [pp 45 and 194]. It has been mentioned that unilateral attacks of Raynaud s phenomenon in a young man may be due to thrombo-angitis obliterans pain in the hand in this disorder may arise from a mild ischaemic neuritis.

Brachial neuralgia from root compression is rarely associated with more than slight oedema and cyanosis of the hand. But vasomotor symptoms—a cold, blue and sometimes swollen hand—are common in the cervical rib syndrome.

A much more difficult problem can be presented by the shoulder hand syndrome When both shoulder and hand show the characteristic picture [p. 84] little or no diagnostic difficulty should arise But it is

thought that on occasion the disorder may be 1 confined to the shoulder in which case there is nothing to distinguish it from acute subacromial bursitis 2 confined to the hand. The skin of the hand is smooth and taut, at first dusky pink or red, later pale and cyanotic the hand and fingers are swollen and painful the fingers held in slight flexion. It is distinguished from rheumatoid arthritis by the much more severe pain, and the much more marked vasomotor changes.



FIG. 33
Tuberculosis of the wrist.

Sudeck s Atrophy This is another form of reflex sympathetic dystrophy which may follow an injury to the wrist tormenting pain affects the whole limb the hand is immobile, oedematous, moist and cyanotic.

Oedeme Bleu des Hysteriques Our attention has been drawn recently[1] to the almost forgotten oedème bleu des hystériques of Charcot, and further instances recorded. In one of these hand and forearm were

[1] Macalpine, I., and Ross, J P (1956), *Lancet* 1, 78.

swollen, skin purplish red, tight and shiny, with several deep ulcers movement and sensation reduced, severe pain at times It is a hysterical manifestation, and completely curable by psychotherapy

PAINFUL HANDS FROM BONE DISEASE

Tuberculosis

Tuberculosis of the Carpus This occurs in children and young people, but is rare. A painful doughy swelling of the wrist appears early and *increases rapidly flexion deformity develops. There is marked wasting* of the forearm and hand muscles. It is unlike an arthritis of the rheumatoid type, in the suggestive large spindle shaped doughy swelling X ray films show severe porosis of the carpal bones, bone destruction, loss of joint space, and eroded articular margins but the diagnosis cannot be made from the X ray appearances alone [FIG 33]

Tuberculous Dactylitis. This condition is a chronic tuberculous osteitis of the phalanges or metacarpals, which is not uncommon in children under the age of 5

Tumours

Enchondroma. An enchondroma or a cyst which is a degenerated enchondroma, in a metacarpal or phalanx, may cause slight aching, but usually is not painful.

Osteoid Osteoma [p 26] This tumour has been recorded in the carpus, metacarpals and phalanges, causing severe pain. Diagnosis can be made only by radiography and subsequent biopsy

Kienbock's disease

This is one of a group of disorders of epiphyses and small bones called osteochondritis juvenilis It affects the lunate bone [FIG. 34]. There is a patchy sclerosis and rarefaction, and the bone becomes deformed The nature of the change is obscure there is commonly a history of trauma, and it has been regarded as an aseptic necrosis. It affects young adults—usually young men. *The complaint is of an aching* wrist. Later osteo-arthritic changes will occur in the radio-carpal joint, and pain is likely to become more severe. The diagnosis can be made only by a radiograph

Madelung s deformity

This is a gradually increasing painful deformity of the wrist that begins in adolescence. It is thought to be a growth defect. The lower end of the radius curves increasingly forwards. The lower end of the ulna does not follow it the head and styloid process are displaced back wards and form a marked prominence on the dorsum of the wrist.



FIG 34
Kienbock's disease
(by kind permission of Dr Cecil Bull).

PAIN IN THE REGION OF THE HIP

The hip in popular speech means vaguely the root of the lower limb A request for localization with more precision may reveal that pain is also in the groin and anterior thigh or that it is confined to the groin, or the trochanteric area, or the buttock It may be that one sided pelvic pain is reported as pain in the hip

What we have to decide is, if the condition is 1 an arthritis of the hip joint, or one of the several disorders that may mimic this or 2. one of the many causes of pain in the groin, trochanteric area or buttock, that do not simulate hip disease.

ARTHRITIS OF THE HIP JOINT

IN CHILDREN

In children arthritis of the hip usually presents itself at the onset as a limp This is why the child is brought for advice he may complain of an ache in the hip region, but not of bad pain. The differential diagnosis of arthritis of the hip in children is therefore considered under limping [p. 64].

IN ADULTS

Pain from the hip joint is felt characteristically in the root of the limb, the groin the anterior thigh and often the knee of the affected side.

If all movements at the hip are restricted, arthritis of the joint is highly probable but not certain. The differential diagnosis is

ARTHRITIS OF THE HIP

osteo-arthritis rheumatoid arthritis ankylosing spondylitis suppurative arthritis tuberculous arthritis

CONDITIONS SIMULATING ARTHRITIS OF THE HIP

Bone disease at the upper end of the femur new growth osteoid osteoma

Osteomalacia

Ilio-psoas bursitis

Psoas abscess

Pelvic disease

X ray films will usually confirm the presence of some form of arthritis of the hip Conditions simulating it are comparatively rare, and they can usually be excluded by the appropriate negative clinical data, particularly those provided by an examination of the inguinal region and pelvis.

Signs confirmatory of arthritis are 1 deformity the lower limb is held flexed at the hip abducted or adducted, and rotated 2. wasting of the gluteal muscles over the affected hip.

Osteo-arthritis of the hip

This affects mainly the middle aged or elderly When seen in younger patients it is likely to be an end result of coxa vara, slipped epiphysis, or Perthes disease in childhood It affects both men and women, being slightly more common in men.

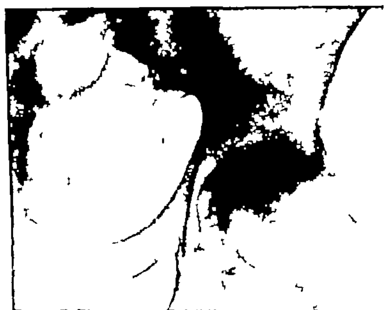
The first complaint is of stiffness he is no longer able to cross one leg over the other in comfort. There may be some pain or aching in the *groin and anterior thigh. Usually pain is quite moderate at first* it may be referred to the knee and not felt in the hip at all for a time.

Early demonstration of restriction of all movements may need very careful comparison with the sound side The range diminishes with advance of the disease, and the final state may approach complete rigidity

Flexion and adduction deformity gradually develop from wasting of the glutei—the extensors and abductors of the hip To keep the normal position of the feet on the ground the pelvis is tilted and there results *an apparent shortening of the affected limb*



(*a*)



(*b*)

FIG. 35
Progressive osteo-arthritis of the hip. (*a*) early (*b*) two years later

X ray appearances are very characteristic. There is narrowing of the joint space in the upper segment of the acetabulum, lipping of the acetabular rim and that of the articular surface of the femoral head. There is an irregular bony sclerosis, side by side with local osteoporosis or even cavity formation The head is often flattened, displaced upwards on the acetabulum and in later stages may be mushroomed by osteophytes [FIG 35]

Rheumatoid arthritis [p 190]

Rheumatoid arthritis affects the hip if at all, late in the disease. It does not begin in the hip and therefore the diagnosis will be well established before this joint is affected.

Ankylosing spondylitis [p 87]

This condition may have an arthritis of the hip as its first manifestation Pain is likely to be severe, and the joint may quickly ankylose. The type case is a young adult male. The X ray appearances are those of rheumatoid arthritis decalcification of articular bone, narrowing of the joint space, and possibly articular erosions

Suppurative arthritis

Suppurative arthritis may be staphylococcal or streptococcal in origin, the organism having invaded the joint from the blood stream, or from a peri-articular infection. The patient is gravely ill, with high fever Pain from the hip is severe restriction of hip movements is pronounced extreme tenderness can be elicited by palpation over the joint and by pressure on the trochanter The hip is held flexed and rotated. The condition is rare in adults more common in infancy and early childhood

CONDITIONS SIMULATING ARTHRITIS

Conditions that may mimic arthritis of the hip are uncommon but some are so serious that in doubtful cases they must be sought with care. A case is doubtful if in adults the X ray films are negative, or if in the elderly they show no more than normal ageing changes in the joint.

In the groin it is possible that ilio-psoas bursitis, or psoas abscess may be detected though both are uncommon [pp 66 136]

Some pelvic diseases in women—salpingo-oophoritis, carcinoma of the uterus, ovarian endometriosis—may give severe pain felt over a wide area the lower abdomen and pelvis, sacrum, iliac crests and groins, down the inner side and back of the thighs. It can be that all movements at a hip joint are restricted by pain, and the condition may strikingly simulate acute arthritis of the hip

Repetition of the X ray examination may reveal a new growth not detected in the first films. Sarcoma at the upper end of the femur or in the pelvic bones, may easily be mistaken for arthritis of the hip—particularly if early X ray changes are missed and the joint happens to show slight degenerative changes.

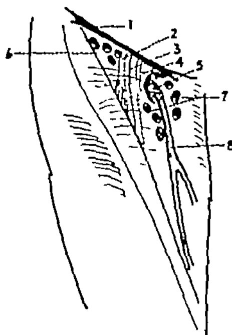


FIG 36

The inguinal lymph glands 1 Inguinal ligament 2. femoral N 3 femoral A 4 femoral V 5 fossa ovalis 6. superficial inguinal glands 7 superficial subinguinal glands 8. great saphenous vein.

Osteomalacia [p 32] This and other general rarefying diseases of bone only mimic disease of the hip joints, in that prominent symptoms are pain in the legs and back, and a waddling gait from spasm of the abductors of the hip The general muscular weakness, lassitude, bone tenderness and deformities, and the appearance of the bone in X ray films, will prompt the special investigations to establish the diagnosis.

PAIN IN THE GROIN

The groin (inguen) strictly means the inguinal region—the part of the abdominal wall just above Poupart s ligament but we may conveniently include the area of Scarpa s triangle—the femoral region

LOCAL LESIONS

Local lesions are often associated with swelling, and I therefore include a brief reference to swellings that are not painful.

Enlarged lymph glands

There are 10-20 glands, in two groups

1 A chain immediately below the inguinal ligament, draining genitals, perineum, buttocks and lower abdominal wall.

2. A group on each side of the upper part of the great saphenous vein, draining the lower limb [FIG 36]

Most of these glands are subcutaneous

An enlarged gland is sometimes difficult to distinguish from an irreducible femoral hernia but a hernia gives one swelling only and it is rare for only one gland to be enlarged.

When enlarged glands are detected in the groin it is important to examine the cervical and axillary regions in case we are dealing with a general lymphadenopathy We need not discuss here the differential diagnosis of this for only glands enlarged from local causes are painful.

Enlarged lymph glands confined to the groin are either

1 due to infection from the drainage area. The tender swollen glands, with reddening of the overlying skin, and the presence of sepsis anywhere in the drainage area, make the diagnosis fairly obvious.

2. malignant. Carcinoma is uncommon but may occur it is secondary to epithelioma of the genitals, perineum, leg or foot. Characteristic are the hardness of the glands, their adhesion to deep fascia and skin and the considerable pain.

Melanotic sarcoma may be large and rapidly growing when the primary growth is very small. Some cases show melanuria (the freshly passed urine has a normal colour but it contains a colourless melanogen which on exposure to the air quickly oxidises to melanin, colouring the urine first brown and eventually black. Immediate blackening of the fresh urine occurs on addition of 1 nitric acid 2. a solution of ferric chloride)

Abscess

An acute abscess in the groin is likely to be due to suppuration of infected lymph glands. A chronic abscess may form in the groin as a result of tuberculosis of 1 glands 2. hip joint 3 sacro-iliac joint, 4 spine (psoas abscess) [p. 66]

Ilio-psoas bursitis

The bursa lies between the tendon of the ilio-psoas and the capsule of the hip joint. Bursitis is not common. It may be confused with

arthritis of the hip because the thigh is held slightly flexed and externally rotated and all passive movements at the hip are slightly restricted by pain.

Sometimes the inflamed bursa is distended when it may be confused with other femoral swellings The bursal swelling is external to the femoral vessels. Swellings in the same position that might therefore be confused with it are psoas abscess [p 66] and lipoma A femoral hernia, and a saphena varix form a swelling internal to the femoral vessels that of an inguinal hernia is above Poupart s ligament.

REFERRED PAIN

An experimental pain stimulus applied to structures supplied by L.1 and L.2 nerve roots evokes pain in the groin and iliac crest areas. Clinically any painful lesion of L.1 or L.2 vertebrae gives such referred pain the spine is therefore always examined clinically and radiologically in unexplained pain in the groin.

The principal visceral disorders associated with referred pain in the groin are

1 renal colic, the pain being felt in the loin and lower abdomen and shooting down to the groin

2 pelvic disease a congestion of the pelvic organs from any cause may give pain radiating widely to the sacrum and iliac crests and groins and lower abdomen.

PAIN IN THE BUTTOCK

The main causes are

A LESION OF THE LUMBO-SACRAL SPINE [p 85]
in young people especially *ankylosing spondylitis tuberculosis of a sacro-iliac joint*

DISEASE OF THE INNOMINATE BONE

A PAINFUL LESION IN THE SOFT TISSUES *fibrositis sacro-iliac strain gluteal bursitis*

THE LUMBO SACRAL SPINE

The gluteal muscles derive their nerve supply from L.4 5 S 1 2. Any painful lesion of the spine involving one or more of these nerve roots will be felt in the buttock. Most of these conditions are described under 'LOW BACK PAIN' [p. 85]

Any type of arthritis of the hip joint gives anterior crural pain. Some weakness of the gluteal muscles of the affected side develops, and often therefore some general aching in the buttock. But pain here, especially posterior crural pain associated with osteo-arthritis of the hip, is nearly always caused by a co-existing lumbar spondylosis

Tuberculosis of a sacro-iliac joint

Tuberculosis of a sacro-iliac joint is comparatively uncommon, being less frequent than tuberculosis of the spine It affects young people, the maximum incidence being 16-35

It is a unilateral disease. The prominent symptom is insidiously increasing pain, felt in one buttock, and often in the back of the thigh and upper part of the calf

The pain may be thought to be sciatic neuralgia but it does not reach the ankle, nor the lateral part of the calf Paraesthesia, loss of ankle jerk, and other neurological signs are absent, except in the very rare instances when a cold abscess *anteriorly has involved the lumbo-sacral cord*

Swelling over the joint may be found. A cold abscess may be the first sign and in any case is likely to appear eventually It usually forms posterior to the joint, but may track to the gluteal fold, the loin, or elsewhere. Sinuses develop in some 50 per cent. of patients.

Tuberculous lesions are likely to be detected elsewhere—e.g. in the lungs or in the spine A careful clinical investigation is essential in all suspected cases.

In a young patient complaining of aching pain in one buttock, of insidious onset but increasing in severity and eventually causing a limp *X ray films of the sacro-iliac joints should be examined with care.* The changes may at first be equivocal, striking and characteristic changes being late in appearing. The only early change may be erosion of the joint surface, perhaps limited to the lower end of the joint, which may appear broader than on the sound side. There may be no local rarefaction, *which makes early diagnosis difficult.* Later erosion becomes deeper there is adjacent osteoporosis, and perhaps the appearance of a sequestrum. There is little or no peripheral sclerosis, as in ankylosing spondylitis moreover the changes of ankylosing spondylitis are always bilateral, those of tuberculosis always unilateral.

Disease of the innominate bone particularly Paget's disease [p. 28], and neoplasms, may be responsible for pain in the buttock.

PAINFUL LESIONS IN THE SOFT TISSUES

Often we cannot get beyond a diagnosis of fibrositis But as this syndrome in some 50 per cent or more of instances is no more than

referred somatic pain and tenderness, the diagnosis should not be accepted until all possible causes of referred pain have been considered and eliminated.

There are two instances of true fibrous tissue inflammation that may be responsible

Sacro-iliac strain

Sprain of the sacro-iliac ligaments may it is generally believed occur during parturition. The symptoms are pain in the buttock and back of the thigh a dull pain, with shooting stabs on jolting or twisting movements. These symptoms, however are also those of any disorder at the lumbo-sacral junction including an intervertebral disc lesion. Confident distinction may be impossible [p. 101]

The symptoms may become chronic.

It was formerly believed that chronic sacro-iliac strain could arise from rapid increase in weight, or unaccustomed prolonged stooping over work of some sort, especially if the ligaments have lost tensile strength through chronic ill-health. But nowadays there is some doubt that the condition exists.

Gluteal bursitis

Of the numerous bursae in the gluteal region, two are important in that local pain may arise from bursitis.

1 *Bursa trochanterica m glutei maximi* is a large bursa constantly found lying on the postero-lateral surface of the great trochanter and origin of the vastus lateralis, and covered by the gluteus maximus close to its insertion. It is liable to tuberculous bursitis, causing local aching and a tender swelling But this is rare

2. *Bursa ischiadica m glutei maximi* lies on the posterior surface of the ischial tuberosity and the origin of the triceps and semitendinosus, and is covered by the lower part of the gluteus maximus It is liable to chronic bursitis, possibly from prolonged sitting on a hard surface.

PAIN IN THE INTERCOSTAL REGION

Pain round the ribs —i.e. in some part of the course of one or more ribs or interspaces, usually the lateral or anterior part—is either root pain, or pain arising in the chest wall or thoracic cavity It is usually on one side of the chest if on both, at the same level it is called a girdle pain

Root pain is continual, with intermittent paroxysms of severe pain, aggravated by coughing or sneezing, or by movements of the spine. Pain arising from the chest wall or thoracic cavity has a more steady quality but is aggravated by deep breathing.

Both clinical and X ray examination of the dorsal spine and chest are essential at times X ray films of the ribs in the painful area should be obtained

The causes of intercostal pain are

ROOT PAIN

likely to be unilateral

dorsal disc protrusion

dorsal disc degeneration

herpes zoster

girdle pain (but unilateral in some instances)

tabes

disease of the vertebrae tuberculosis metastatic carcinoma myelomatous ankylosing spondylitis

extramedullary tumours

syphilitic pachymeningitis

intrathoracic aneurysm or new growth.

CHEST WALL

disease of the ribs

fibrositis

LUNGS AND PLEURA

See dorsal pain [p. 108]

ROOT PAIN

Dorsal disc protrusion

We know we are dealing with intercostal root pain from dorsal disc protrusion only in those instances associated with signs of cord compression for these are the only cases operated on. Very few such operations have had to be performed In recorded instances severe unilateral root pain was the presenting symptom and this was followed by progressive spastic paralysis of the legs and other signs of cord compression

It seems probable, however that dorsal disc protrusions causing root pain only are quite common but there are no positive diagnostic signs, and diagnosis rests on the clinical picture and exclusion of other possibilities

There is a sudden seizure of severe pain on one side of the dorsal spine, radiating round the ribs on that side It has the characters of root pain. It usually abates in a week or so but may recur and some endure

for many weeks. X ray films of the dorsal spine are likely to show no abnormality

Herpes zoster

This condition is peculiarly common in one or more of the thoracic segments, usually unilateral In middle life and after the rash is preceded by 3 or 4 days of intercostal pain When the rash appears the diagnosis is obvious before it is not possible Therefore it is unwise to commit oneself during the first few days of any intercostal neuralgia.

After the rash has faded, a most severe and intractable neuralgia may persist in elderly patients.

Girdle pain

Bilateral root pain gives a sensation of a painful tight band or belt encircling the trunk. There is a girdle sensation of different origin being the highest limit of sensory loss, or of spasticity in paraplegia due to a dorsal cord lesion, as in disseminated sclerosis.

The main groups of causes of girdle pain are

1 *Tabes* in which it is a common early symptom [p. 165]

2. *Diseases of the spine* those likely to cause bilateral root pain are those leading to collapse of a vertebral body notably tuberculosis, metastatic carcinoma, and myeloma it is not uncommon in ankylosing spondylitis [p. 87]

3 *Compression by a cord tumour* [pp 76, 213]. An extramedullary tumour may for many months cause no more than an intractable inter costal neuralgia Its chronicity should make us suspicious and on the alert for evidence of cord compression—sensory changes below the painful level and pyramidal signs in the lower limbs. It is said that a unilateral intercostal neuralgia becoming later a girdle pain is, in the absence of disease of the vertebrae, pathognomonic of extramedullary tumour

Rarer causes are *syphilitic pachymeningitis* and erosion of the spine by mediastinal new growth or aortic aneurysm.

PAIN FROM THE CHEST WALL AND THORACIC CAVITY

When the pain is not suggestive of root pain, the problem is one of distinguishing pain arising in the chest wall from pain due to intra thoracic disease the commonest problem being to distinguish acute fibrositis from acute pleurisy

Pleural pain

Acute Pleurisy This causes a stitch-like stabbing intercostal pain and, as in both neuralgia and fibrositis, the intercostal spaces in the painful area are tender The common mistake is to diagnose pleurisy

when the condition is really intercostal neuralgia or fibrositis. Pain aggravated by deep breathing is common to all But a patient with pleurisy is febrile has an ineffective and painful cough, and a pleural rub will be heard over the painful area

Chronic Pleurisy Chronic pleurisy and pleural adhesions may be a cause of pain in the chest, as may many other intrathoracic diseases [p. 109].

Fibrositis

Acute fibrositis is often a wrong diagnosis for referred pain from the spine. A drop or two of strong saline injected into an intervertebral ligament of the upper dorsal spine will evoke a steady pain in the corresponding intercostal space lasting several minutes The responsible lesion may be *disc degeneration sprain of a deep spinal ligament* or *chronic postural strain*

But I believe that sometimes the primary condition is a localized myalgia which we may still call fibrositis

Epidemic myalgia (BORNHOLM DISEASE DEVIL'S GRIP)

This is the only proved instance of myalgia due to virus infection There is a sudden onset of pain, from what appears to be a myositis of the diaphragm in the region of its insertion. There is intermittent pyrexia the total duration does not exceed 14 days. Its severity varies from malaise and pain on deep respiration, to sudden paroxysmal pain with high fever and abdominal rigidity

It is possible that in many instances acute fibrositis abating in 10 days or so coming in small epidemics, is due to a similar myotropic virus. But this is not proven.

Tietze's disease

This painful tender swelling in costal cartilages may be mentioned here It was first described in 1921 but has been noted in this country only during the past 15 years. Its onset is insidious aetiology and pathology quite obscure. It affects one or more costal cartilages, usually *the second X ray films are taken to exclude intrathoracic disease*, or disease of the rib The swelling eventually subsides.

PAIN AND INTERMITTENT CLAUDICATION

If a patient has powers of clear description, the symptom intermittent claudication will be recognized at once. But too often the complaint is just of pain in one calf sometimes in one foot, or occasionally in one hip or thigh, coming on while walking.

The characteristic features of this peculiar pain are that it never occurs at rest or when standing still it only comes on after having walked a certain distance. A cramp-like pain gains in intensity so quickly that the patient must stop and having stopped although still weight bearing, relief is complete often within a few seconds. When he walks on again pain returns after the same distance, and so on with a regular periodicity

The cause of the pain is muscular exercise under ischaemic conditions. A reduced blood flow to the limb may be adequate for a resting muscle but gravely inadequate for the muscle during exercise Repeated con tractions of an ischaemic muscle lead to accumulations of pain-producing metabolites, and pain ceases only when a more adequate circulation is restored by rest.

The circulatory defect is nearly always due to *occlusive arterial disease* Naturally the amount of exercise possible varies with the degree of ischaemia. When the blood flow is but slightly reduced the patient may be able to walk for half an hour or more and the pain that comes on may be relieved by standing still for a few seconds. But the greater the reduction the shorter the distance he can walk. It may be a few yards only and when he stops pain may last for many minutes. Thus from the patient s account we may roughly gauge the degree of arterial occlusion.

Pain is usually unilateral even if the vessels of both legs are diseased This merely indicates an unequal degree of occlusion the patient is halted by pain in the more ischaemic limb before the limb with the more adequate blood flow is affected

PHYSICAL SIGNS OF OCCLUSIVE ARTERIAL DISEASE

The first decision to be made is that the pain is in fact intermittent claudication For if from the patient s description this is not clearly so we may be dealing with pain from, for example, sciatica, pes valgus, or osteo-arthritis of the hip It is necessary therefore to try to demonstrate a defective circulation in the limb The most important signs are

Absent arterial pulsation

The most helpful clinical examination is palpation of pulses. Most claudication patients have no pulse below the femoral if pulsation is normal, pain in the limb is probably not of this nature. Partial obstruction of an artery will abolish the pulse below it, although not arresting the blood flow

The pulses to be palpated are

femoral with the patient recumbent, palpate just below Poupart s

ligament with the tips of the index, middle and ring fingers in line with the ligament.

popliteal with the patient prone, one hand flexes his leg at the knee, and the thumb of the other palpates

posterior tibial the artery runs just distal and posterior to the medial malleolus supporting the patient s heel with one hand, place the other over the front of his ankle and palpate with the finger tips.

dorsalis pedis transfer the finger tips to the line of the dorsalis pedis artery where it runs down the dorsum of the foot, passing usually between the bases of the first and second metatarsals, or it may be lateral to this. Dorsalis pedis pulsation cannot be felt in 10 per cent. of normal people.

Skin temperature and colour changes

The colour of the feet may be normal. But if arterial disease is at all advanced, the foot is red and cold and it may be mottled with dusky and blanched areas if the circulation is very slow or absent.

Postural colour changes—abnormal pallor on elevation, and dependent rubor—are an important indication of impaired circulation.

If the feet of a healthy recumbent person are raised, and held up for about 2 minutes, a mild pallor will be observed But if occlusive arterial disease is present in the lower limbs, the pallor is much more marked and may reach an extreme degree.

If this is observed and especially if it is asymmetrical, ask the patient to sit with his legs hanging over the edge of the couch. With normal arteries the colour returns in about 15 seconds. If it does not, there is some arterial deficiency if normal colour has not returned in 30 seconds, impairment is marked and is extreme if the foot is still pale after 60 seconds.

With continued dependency and return of colour excessive rubor develops in occlusive arterial disease.

When the impairment of circulation is asymmetrical, the foot of the more severely affected limb will be obviously colder than the other simple palpation will detect it. While coldness of both feet is of little or no diagnostic value, one foot obviously colder than the other is very suggestive of occlusive arterial disease.

DIFFERENTIAL DIAGNOSIS

Having determined that the complaint is in fact intermittent claudication it remains to determine the cause of the reduced blood flow The vessels affected are the main arteries of the limb

Arteriosclerosis Obliterans In women, and in all patients over 50, the

disease is most probably arteriosclerosis obliterans. Symptoms are practically confined to th lower extremities. Men are more often affected than women the maximum incidence is 50-70 years Hyper tension is found in 35 per cent —an incidence probably not greater than among patients without arteriosclerosis Claudication may for many years be the only symptom but there may be others A sensation of coldness in one limb is often an early symptom Numb tingling or burning paraesthesia in the feet is common Severe ischaemia may cause rest pain—sleep is disturbed by a dull aching pain in the toes or foot. It may also cause trophic changes atrophy of the subcutaneous fat and muscles of the leg atrophy of the toes and nails ulceration and gangrene, often first appearing round the nails osteoporosis of the bones of the foot. In these advanced cases there is likely to be stiffness of the joints, and oedema of the feet from prolonged inactivity the patient walks so little that claudication will not be his presenting symptom

Thrombo-angiitis Obliterans In young men the disease is more likely to be thrombo-angiitis obliterans. This disease affects males almost exclusively and the maximum incidence is 25-40 years. Slightly more common among Jews, it affects men of any race The vessels most commonly diseased are the anterior and posterior tibial and the radial and ulnar arteries. The symptoms and signs, being those of ischaemia, are the same as those of arteriosclerosis But an important distinction is that thrombo-angiitis is an obliterative disease of veins as well as arteries. A thrombophlebitis migrans affecting the superficial veins may be the first manifestation of the disease Red, indurated tender cords up to about one inch in length may be found anywhere on the upper or lower limbs.

The important clinical features distinguishing arteriosclerosis from thrombo-angiitis obliterans are summarized in the Table

	ARTERIOSCLEROSIS	THROMBO-ANGIITIS
maximum incidence	50-70 years	25-40 years
sex	M F 6 1	almost always M
upper extremities	rarely affected	often affected
veins	never affected	superficial thrombo-phlebitis migrans common
calcification of arteries in radiographs	common	none

Causes of claudication other than arteriosclerosis and thrombo-angiitis are rarities. It may be a sequel of sudden arterial occlusion from

an embolus, or thrombosis, the collateral circulation being inadequate. It may be a result of aneurysm of the main artery or of an arterio-venous fistula.

PAIN IN THE KNEE

SWELLINGS IN THE VICINITY OF THE KNEE

A swelling in the popliteal region is nearly always either a Baker's cyst or a distended semimembranosus bursa. Aneurysm of the popliteal artery abscess, enlarged glands, and a soft tissue tumour are possible but rare. A primary new growth of bone [pp 21-6] in the upper end of the tibia or the lower end of the femur may cause a swelling in any situation round the knee.

The commonest swelling in front of the knee is a pre-patellar bursitis. Osteochondritis of the tibial tubercle is a not uncommon cause in children and adolescents.

At one side or the other the only common swelling is a cartilage cyst. Here a lipoma is a possibility

Popliteal swellings

The commonest popliteal swelling is *Baker's cyst* First described some 80 years ago, it is a posterior herniation of the synovial membrane of the knee, through a weak part of the capsule. Passing between the two heads of the gastrocnemius, it forms a tense rounded swelling in the lower central part of the popliteal space It is usually associated with arthritis of the knee joint, but may occur independently

The *semimembranosus bursa* is between the semimembranosus and the medial head of the gastrocnemius it is prolonged onto the capsule of the knee, and often communicates with the joint. Bursitis is common at any age, often symptomless in young adults, often associated with osteo-arthritis of the knee.

With the leg extended there is a tense swelling in the medial part of the popliteal space.

It is unlikely that these two fairly common conditions would be confused with solid soft tissue popliteal swellings and these—enlarged lymph glands, lipoma, sarcoma, are very rare. Their features also are quite different from those of acute popliteal abscess.

They might be confused with *popliteal aneurysm*. But this is a compressible tumour that pulsates, with an expansile pulsation synchronous with the heart beat. With a stethoscope a bruit may be heard over it.

The complaint is of pain in the knee and leg. Its pressure on the popliteal vein causes varicosity of the veins of the leg.

Anterior swellings

Pre-patellar Bursitis (Housemaid's Knee) This is easily recognized the enlarged bursa forming a distended sac over the lower part of the patella. Trauma is probably the commonest cause. But an infective bursitis is fairly common sometimes associated with secondary effusion in the joint.



FIG. 37

Osgood Schlatter disease (osteochondritis of the tibial tubercle) (by kind permission of Dr Cecil Bull).

Osgood-Schlatter Disease (Osteochondritis of the Tibial Tubercle) This presents as a swelling, usually painful and tender in the region of the tibial tubercle, in a boy—less often a girl—of 10 to 15 years. Lateral X ray films show an irregular tibial tubercle, with areas of decalcification [FIG 37] These appearances without local swelling may be of no significance

Medial and lateral swellings

Nearly all of these swellings are semilunar cartilage cysts. These multilocular cysts form in the periphery of the inter-articular cartilage especially the external cartilage. They are within the joint capsule. The presenting symptom may be a lump, or a painful knee On examination,

a swelling is found at the side of the knee at the joint level, the size varying from one just palpable to a large tense lump

SYNOVIAL EFFUSIONS IN THE KNEE

RECURRENT EFFUSIONS

The main causes are

Cartilage injury Loose body Osteochondritis dissecans Intermittent hydrarthrosis and palindromic rheumatism Chondromalacia patellae Brodie s abscess

Semilunar cartilage injury

This results from severe rotation strain at the knee at a moment when the limb bears the weight, and the knee is slightly flexed The internal cartilage is the one most commonly injured it is torn from its capsular attachment, crushed between the articular surfaces, and perhaps split in its length [FIG 38]

There is a sudden severe pain at the inner side of the knee, and the limb cannot be fully extended at the joint. This locking is the classical sign repeated effort may end in a sudden yielding of the obstruction, or full extension is gradually regained. Within a few hours synovial effusion appears With rest, pain and effusion subside after some 3-4 weeks, and the joint may then seem more or less normal again.

It is, however prone to recurrent attacks of pain and effusion, often from quite trivial strains or for no apparent reason.

Diagnosis soon after the injury can be made with confidence if there is locking, and localized tenderness anterior to the internal lateral ligament in the joint line and if X ray films exclude the presence of a loose body A source of error is that the torsion strain may have merely sprained the attachment of the cartilage to the tibia, without causing a cartilage injury pain, effusion, and tenderness in the joint line may follow but there is no locking.

Diagnosis may be difficult in recurrent attacks unless there is a typical history

But a cartilage injury is often not typical in its clinical signs. Tears of the free margin, or far back in the posterior horn, or of the external cartilage do not cause locking.

Attacks of pain and effusion may recur for years, and the history will be merely of a wrench of the knee followed by severe pain and swelling. These less common injuries can be recognized by *McMurray s sign*, if it is present. The patient lies on his back with the knee fully flexed. Passively extend the leg on the thigh with the leg held externally rotated

on the thigh an audible click or clunk during the movement indicates a lesion of the posterior part of the internal cartilage Repeat, if necessary holding the leg internally rotated on the thigh the same sign indicates a lesion of the posterior part of the external cartilage

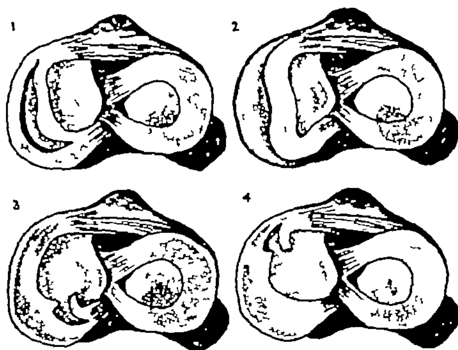


FIG 38

Various injuries of the internal semilunar cartilage 1 bucket handle tear 2. displacement of the cartilage 3 tag of posterior horn 4 tag of anterior horn.

Loose bodies

Among the less obvious forms of trauma is that due to nipping of a loose body

Loose bodies in the synovial cavity may be found in association with osteo-arthritis either as a result of the joint disease, an osteophyte having broken off or as the primary disorder

Two rare causes of loose body are

Osteochondritis Dissecans This is a local necrosis leading to separation of a piece of cartilage and underlying bone from the articular surface of the medial condyle of the femur [FIG 39] or from the articular surface of the patella. The piece drops out from its deep bed. The loose body is single, or at most there are two

Synovial Chondromatosis. This is a benign tumour formation, in which very numerous cartilaginous bodies with calcified centres, are

either free in the joint cavity or attached by pedicles to the synovial membrane. Many hundreds have been found in one knee joint.

Another type of loose body is the melon seed body found not only in joints but in tendon sheaths and bursae They are composed of concentric layers of fibrous tissue little is known of their origin.

When a loose body is nipped between adjacent bones there is a sudden locking of the joint, followed by synovial effusion.



FIG 39
Osteochondritis dessecans
(by kind permission of Dr G A. S. Lloyd).

Intermittent hydrarthrosis

This name is given not to any recurring synovial effusion in the knee, but to one with very special and distinctive characters. It is a rare condition. The incidence is mainly in the 3rd and 4th decades, equally in the two sexes.

Periodically the knee is subject to a rapidly increasing effusion,

reaching its maximum in 2 or 3 days, stretching the capsule taut as a drum, then as quickly subsiding till after about a week from the onset the joint is apparently normal again. Pain is often severe, though some times quite moderate. Attacks occur with a definite periodicity the intervals of freedom, in which the joint seems entirely normal, are often from one to three weeks, but may be a month or even three months. This regular periodicity is the cardinal feature. A. E. Garrod, who reviewed 70 instances of this phenomenon in 1910 wrote that only those cases in which there is a regular periodicity are to be classed as examples of intermittent hydrarthrosis

Palindromic Rheumatism. This condition is similar but there is no regular periodicity and any joint may be affected. Usually only a single joint is involved in each attack. Considerable effusion and pain reach a maximum in a few hours to a few days, and as quickly subside. Even when hundreds of attacks have occurred over many years (I have seen one case of over 20 years duration), there is no residual joint damage. Attacks are afebrile E.S.R. and radiographs are normal. Some cases so diagnosed however have developed rheumatoid arthritis after a variable time. There is some doubt about the relation between the two conditions.

Chondromalacia patellae

This is a name given by orthopaedic surgeons to early osteo-arthritis in the patello-femoral component of the knee joint, at a stage when the only change is degeneration of the articular cartilage, and reactive changes in the subarticular bone of the patella.

It may be responsible for a recurring painful effusion in the knee and can be recognized only by lateral X ray films of the joint. The early X ray changes are areas of rarefaction and sclerosis in the subarticular bone of the patella later there are the typical appearances of osteo-arthritis in the patello-femoral component.

Brodie s abscess

This is described under bone swellings (p 21). It is a cause of recurrent effusions with aching and stiffness of the joint.

CHRONIC EFFUSIONS

The main causes are

TRAUMATIC SYNOVITIS
- contusions sprains quadriceps weakness

OSTEO-ARTHRITIS

ARTHRITIS
- rheumatoid tuberculous gonococcal Reiter s disease syphilitic synovial reaction to extra articular inflammation

Traumatic synovitis

Bone injury having been excluded by radiography the only diagnostic problem is to determine if there has been an injury to one of the ligaments or to a semilunar cartilage

Sprain of the Internal Lateral Ligament This is a fairly common sports injury resulting from an abduction strain There is a synovial effusion of variable degree and the limb is held in slight flexion at the knee passive extension is painful and there is a point of severe tenderness over the insertion of the upper end of the ligament an inch or so above the joint line. Like any other sprain symptoms may become chronic, effusion, pain, and weakness of the knee persisting for many months.

A hyperextension injury may sprain the *anterior cruciate ligament* As a rule no tenderness can be found but passive extension and antero-posterior movements are painful Again, pain and effusion may become chronic.

Quadriceps Weakness With rest a traumatic effusion should be absorbed in 1 3 weeks but if during this time quadriceps exercises are not carried out regularly and efficiently a weak and wasted quadriceps may be left for many months. From this cause alone the knee is left unstable, and prone to persisting synovitis from repeated minor ligamentous sprains.

Osteo-arthritis of the knee

Synovial effusion if there is any is only slight or very moderate in amount. The symptoms are a feeling of stiffness in the knee after sitting, pain on going downstairs, and it may be pain when standing or walking. In early cases there may be very little to be found beyond wasting of the quadriceps, a harsh crepitus on movement (but this sign is of very little value) and some restriction of flexion and extension of the knee.

If the joint is swollen—as it may be from effusion, and in advanced cases from osteophytes—the swelling is very different from that of a true arthritis of the knee. In rheumatoid arthritis, or any form of chronic infective arthritis, the swelling is predominantly a capsular swelling even with no demonstrable effusion, the suprapatellar prolongation of the capsule bulges on each side imparting to the touch the impression of a soft thickening, or of a semi-solid content. This capsular and synovial swelling, with perhaps a gelatinous structureless material in the joint cavity is never found in osteo-arthritis.

X ray films establish the diagnosis in early cases, the usual even contour at the articular margins of femur tibia and patella has become pointed in places, indicating osteophyte formation and there is

increased density of the articular lamella. In advanced cases there is extensive osteophyte formation the joint space is asymmetrically diminished and loose bodies may be seen [FIG 40]

A diagnosis of osteo-arthritis of the knee is not possible with normal X ray films.



FIG 40
Osteo-arthritis of the knee.

Arthritis of the knee

When effusion is not intermittent it may at times be difficult to know if we are dealing with a traumatic synovitis or an arthritis of the joint. The more intractable it is to the usual physical treatment, the more likely it is to be an arthritis of the knee. Cytological examination of the joint fluid obtained by aspiration may be helpful. Mechanical injury and osteo-arthritis provoke a synovial transudate a fluid of low protein

content and low total nucleated cell count—usually well below 1,000 per c.mm Any non-traumatic inflammation of the synovial membrane causes a synovial exudate, a fluid of high protein content and high total cell count—usually above 10,000 per c.mm. But equivocal counts are not uncommon.

One knee may be the only joint showing signs of arthritis at the onset of *rheumatoid arthritis* [FIG 41] The joint shows some flexion deformity and restricted flexion It is enlarged from capsular swelling and some effusion, with capsular bulges on each side. X ray films show rarefaction of the articular bone ends perhaps nothing else abnormal, or perhaps irregularity of the articular line from erosions. The E.S R. is increased. Unfortunately this picture is not diagnostic of the rheumatoid type of arthritis. The condition may remain monarticular for many months, but arthritic signs in another joint usually appear within a year if it is of the rheumatoid type

If it remains monarticular one should consider whether it may not be *tuberculous arthritis* In a child, adolescent or young adult, there may of course be very little doubt a swollen knee very moderately painful, with smooth even contours, accentuated by marked muscle wasting, the swelling having the characteristic boggy feel from thickening of synovial membrane, capsule and peri-articular structures, at once suggests tuberculosis. But in adults, at least in the earlier stages, it may be clinically indistinguishable from an arthritis of the rheumatoid type. X ray films will not help, if the only change is rarefaction of the articular bone ends [FIG. 42]. The E.S R. is increased in both. A negative Mantoux test practically excludes tuberculosis a positive result is equivocal. The tubercle bacillus can be demonstrated in the joint fluid by guinea-pig inoculation in some 80 per cent. of instances.

It may be impossible to distinguish the two in any way other than biopsy either of the synovial membrane, or of an enlarged inguinal gland should one be found. Biopsy of synovial membrane is simple and safe, and should probably be carried out earlier than is usual. The material is examined histologically and by culture, and by guinea-pig inoculation. The tuberculous or non-tuberculous nature of the arthritis can be established decisively

Other types of chronic monarticular arthritis of the knee are rare.

After *gonorrhoea*—it may be many years after—a chronic synovitis of the knee may occur Swelling may be considerable, from synovial and peri-articular effusion and muscle atrophy is apt to be severe.

It is now believed that most of these cases are instances of *Reiter's disease* venereal in origin [p. 207] A diagnosis of gonococcal arthritis should not be made unless the gonococcus is demonstrated in the





FIG. 41
Advanced rheumatoid arthritis of the knee.

synovial effusion or unless the joint inflammation responds promptly to penicillin.

Syphilitic arthritis occurs 1 as a manifestation of congenital syphilis in children 2. rarely in the secondary stage in adults, as a painless synovitis 3 also rarely in the tertiary stage a granulomatous infective arthritis, with thickened synovial membrane, capsule and peri-articular tissues from gummatous infiltration

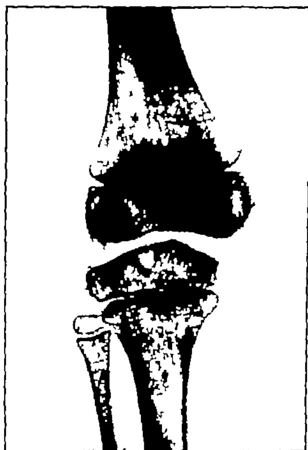


FIG. 42

Tuberculosis of the knee 2 years 3 months after the onset slight osteoporosis, without erosion or loss of cartilage.

The *Charcot joint* results from the sensory disturbance of tabes. It usually affects a joint of the lower extremity—hip knee or ankle. There is a large painless effusion, with hypermobility of the joint. X ray films reveal extreme destruction of articular bone, and luxurious osteophyte formation.

MISCELLANEOUS CAUSES OF PAIN IN THE KNEE

The more important causes of painful knees are discussed in the two preceding sections. But if a case appears to be due to none of these conditions, we may have to consider 1 a number of local causes, mainly rare 2. pain referred from the hip joint.

LOCAL CAUSES

Disease of the articular bone

primary malignant tumours osteoid osteoma osteomyelitis

Chronic villous arthrosis

Haemarthrosis

Pellegrini–Stieda s disease

Pyogenic infections of the knee

REFERRED PAIN FROM THE HIP JOINT

Arthritis of the hip [p 131]

LOCAL CAUSES

Disease of the articular bone

Primary Malignant Tumours The malignant tumours appearing near the knee are

1 *Osteogenic sarcoma* which occurs in young people who complain of increasingly severe pain soon followed by a swelling.

2. *Giant cell tumour* (*Osteoclastoma*) which also occurs in young people. It is only locally malignant and produces a painful swelling, but the pain is less severe than that of a sarcoma.

Osteoid Osteoma [p. 26]. This is a rare benign tumour occurring in adolescents and young adults. Commonest in a long bone of the lower limb it may occur near the knee, and may cause a synovitis. The prominent symptom is persistent localized pain and tenderness.

A radiograph establishes at least the probability of a bone tumour

Acute Osteomyelitis This, although any bone may be affected is most common near the knee. The infection at first is almost always localized in the metaphysis, and spreads to the adjacent periosteum. It is a disease of childhood, affecting boys more often than girls. When, as is very often the case infection starts in the upper tibial metaphysis, there is severe pain and tenderness near the knee, of fairly sudden onset. Tenderness is most severe over the metaphysis Febrile symptoms vary with the virulence of the infection the temperature may rise to 103° F or more.

Later the tissues become swollen, red and oedematous but at least a provisional diagnosis should be made before this

The differential diagnosis is from rheumatic fever and the most helpful criteria are

1 pain remains in one joint a febrile illness cannot be rheumatic fever if pain is confined to one joint

2. there is marked localized tenderness over the infected bone, rather than the general tenderness of a rheumatic joint.

In acute osteomyelitis there is a leucocytosis, often in the region of 25 000 per c.mm but the count is not always higher than in acute rheumatism, and has therefore little diagnostic value.

Chronic Osteomyelitis This is usually a sequel of the acute infection. There is a form in which a chronic discharge through a sinus persists for years, and which depends for its chronicity on the presence of a sequestrum. Another form, and one likely to cause trouble in diagnosis, is a latent infection which flares up at intervals of months or years, and in which there is no sinus. X ray films show circumscribed areas of osteoporosis, but no sequestrum. During an active phase there is complaint of local aching, swelling and tenderness, which is accompanied by a moderate pyrexia.

Brodie's Abscess [p 21] This is a localized bone infection, by an organism of low virulence, which is chronic from the onset.

Chronic villous arthrosis

Perhaps better called menopausal arthrosis of the knees, this condition is almost confined to women at the menopause.

Both knees ache, are sometimes painful and feel stiff when rising from a chair or from bed in the morning These are the symptoms of early osteo-arthritis—but X ray films of the knees are normal, or show only minimal degenerative changes consistent with age.

On examination there is excessive and often tender fat about the knees There is no synovial effusion, nor capsular thickening movement is not restricted There is often the localized tenderness indicating chronic strain of the internal lateral ligament. Some degree of pes valgus is a very common association.

If untreated, osteo-arthritic changes develop after a few years if properly treated, most of these menopausal knees get well.

Any views on the nature of this condition are speculative. There is evidence that in some instances there is a great increase in the size and number of the normal fatty synovial fringes and villi—hence the name 'villous arthrosis But it seems probable that in addition to the local endocrine disturbance of fat, there is also a cause in the chronic strain

from the general laxity and weakness of ligaments and muscles common at the menopause.

Haemarthrosis

Bleeding into the knee joint may follow severe injury Whereas a traumatic synovial effusion begins some 5 or 6 hours after the injury bleeding into the joint is obvious within half an hour Moreover the consistency of the swelling on palpation appears much firmer than that of a synovial effusion.

Bleeding into the knee may follow trivial injury in *haemophilia* Periodic attacks in the same joint are common the blood each time being absorbed to leave a fairly normal joint but eventually absorption is incomplete, and degenerative changes appear in the joint.

It occurs only in males. The history will reveal excessive bleeding from very slight wounds from infancy The coagulation time is greatly increased.

Pellegrini–Stieda's disease

This is a sequel to a blow on the inner side of the knee, which has led to the formation of a plaque of bone over the medial condyle of the femur It is diagnosed radiologically The symptoms are those of a chronic sprain—pain on the inner side of the knee and local tenderness.

Suppurative arthritis

Pyogenic infection of a joint may occur during the course of many infective disorders, local or systemic otitis media, tonsillitis, pneumonia, osteomyelitis, boils, puerperal sepsis, typhoid fever meningococcal meningitis, gonorrhoea and many others. The organism may of course be introduced directly into the joint by perforating wounds, or by intra-articular injection.

The onset of pain is abrupt, and the joint soon becomes hot, red, swollen and tender A high intermittent pyrexia develops. Movement at the joint is greatly restricted by pain and spasm. With some infections several joints may be affected but a suppurative arthritis is commonly monarticular affecting one of the larger joints.

Aspiration, for cytological and cultural examination of the joint fluid, will establish the diagnosis. There is a high polymorphonuclear leucocytosis.

The commonest infecting organism is *Staphylococcus aureus* or *Streptococcus haemolyticus* Less common joint infections are

Pneumococcal Arthritis This may complicate pneumonia, or be a primary joint infection in children.

Acute Gonococcal Arthritis This may occur 2 3 weeks after the onset

and calf muscles of the affected limb can often be observed In some instances there is weakness of the dorsiflexors of the foot, with slight foot drop

The cause of sciatic neuralgia is injury to the sciatic nerve anywhere in its course in the great majority of instances the lesion is one compressing L.5 or S 1 nerve root as it emerges from its foramen.

The sciatic nerve is derived from L.4-S.3 nerve roots, which arise from the cord opposite T 12 and L.1 vertebrae and descend in the cauda equina to their foramina. The anterior divisions converge and join to form a flattened band on the side of the pelvis, from which it emerges in the lower part of the great sciatic foramen below the pyriformis muscle, as the sciatic nerve This passes between the tuberosity of the ischium and the great trochanter and descends to a point above the knee where it divides into the *tibial* and common peroneal nerves.

The causes of sciatic neuralgia are

CAUDA EQUINA

Compression by meningiomata or other tumours or by gross disc protrusion

INTERVERTEBRAL FORAMINA

Posterior protrusion of the disc L.4-5 or L.5-S 1

Compression of nerve roots by osteo-arthritis

Root compression secondary to disease of the lumbar spine metastatic carcinoma tuberculosis ankylosing spondylitis spondylolisthesis

SACRO-ILIAC JOINT

tuberculosis of a sacro-iliac joint

PELVIC DISEASE

neoplasms chronic inflammation pressure of the foetal head

SCIATIC NERVE

trauma (gunshot wounds and other injuries)

ischaemia of the sciatic nerve } speculative
sciatic neuritis }

It will be obvious that it is in all cases essential to examine the lumbar spine, both clinically and radiologically

CASES WITH LUMBAR SIGNS

Sciatica in a patient in whom there is known to be, or to have been, a primary carcinoma, should arouse suspicion of secondary deposits in the lower lumbar vertebrae. If normal, X ray films should be repeated at intervals in any case of intractable pain [p 93]

Sciatica in adolescents and young adults is equally ominous until

proved not to be due to tuberculosis, either of the lumbo-sacral spine or a sacro-iliac joint [pp 91 138] or to ankylosing spondylitis [p 87]. A true sciatic neuralgia may occur in spondylolisthesis, but chronic backache is the usual symptom [p 101]

Intervertebral Disc Protrusion. If these diseases are excluded, the probable cause of any case presenting well-marked lumbar signs is

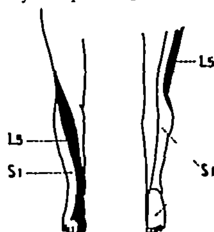


FIG. 43

Approximate area of dermatomes L.5 and S.1 in the foot and leg.

intervertebral disc protrusion—a posterior protrusion of the L.4-5 or L.5-S 1 disc. Disc lumbago [p 86] usually immediately precedes the onset of sciatic pain. Less commonly there is an interval between the attack of lumbago and that of sciatica. Many patients give a history of several attacks of this low back pain, over a period of months or years.

Lumbar signs that suggest disc protrusion are

1 *Sciatic scoliosis* the lumbar spine is held flexed and pulled over to one side by reflex muscle spasm.

2. *Bechterew's sign* the patient cannot lie flat on his back with both lower limbs extended without increased pain relief is obtained by flexing either hip The reason is that flattening of the lumbar curve is protective, and cannot be maintained lying flat with both hips extended

3 For the same reason the patient cannot lie prone.

4 Pressure on the 4th or 5th lumbar spinous process is locally painful, and may evoke a stab of pain in the affected limb

A lateral protrusion of the L.4-5 disc compresses L.5 nerve root. A lateral protrusion of the L.5-S 1 disc compresses as a rule S 1 nerve root but a large one may compress both.

Neurological signs in the painful limb suggesting compression of nerve roots L.5 and S.1 are these

Nerve root L.5 1 Sensory disturbance—impairment or loss of pain sensation or paraesthesia—in a band extending over the antero-lateral aspect of the leg to the medial border of the foot and the 1st and 2nd toes The dermatome of L.5 is shown in FIG 43

2. Weakness of dorsiflexion of the big toe and foot. Among muscles supplied by L.5 are the tibialis anterior and the extensors of the toes [p 13]

Nerve root S 1 1 Loss or diminution of the ankle jerk. The integrity of S 1 is necessary for this reflex.

2 Sensory changes in a band below the knee covering the lateral aspect of the calf the lateral side of the foot, and the sole the dermatome of S 1 is shown in FIG 43

3 Weakness of plantar flexion of the big toe and foot, and of the gluteal muscles of the affected side among the muscles supplied by S 1 are the plantar flexors of the big toe and foot, and the gluteus maximus [p 13]

Pressure on the Cauda Equina This is a rare surgical emergency It may be due to a massive disc protrusion, or to a tumour The signs are paralysis of one or other large muscle groups in the painful limb, and disturbance of the bladder sphincter Marked muscular weakness and atrophy should always excite suspicion that we are not dealing with a simple sciatica.

CASES WITHOUT LUMBAR SIGNS

There is no abnormal posture no awakening of pain on attempted lumbar extension or by pressure on L.4 or L.5 spinous processes. The patient can lie prone, or flat on his back, and turn over with ease. These cases are not common, but by no means rare.

Usually we are left speculating as to the cause it may be a root *compression, giving such slight* lumbar signs that we have missed them it may be an ischaemia of the sciatic nerve or even a sciatic neuritis.

But what in all these cases we must make every effort to exclude is intra pelvic disease, especially if pain is intractable and severe. The disorder that could be responsible is an inflammatory or neoplastic mass compressing the lumbo-sacral plexus.

Femoral neuralgia

Femoral neuralgia is not common—not nearly so common as sciatic neuralgia. There is paroxysmal pain in the front of the thigh, accompanied by some weakness and wasting of the quadriceps, and absent or diminished knee jerk.

The femoral nerve is derived from L.2, 3 and 4 nerve roots, which

emerge from the cord opposite T 10 and 11 vertebrae, and descend with the cauda equina to their respective foramina.

In the lumbar region 95 per cent. of disc protrusions that cause symptoms are distributed equally between the discs L.4-5 and L.5-S 1 The remaining 5 per cent. affect L.3-4 This uncommon disc protrusion compressing L.4 nerve root, is probably the commonest cause of femoral neuralgia.

Some instances may be due to osteo-arthritis of the upper lumbar spine.

There are more serious possibilities diseases of the lower thoracic and upper lumbar vertebrae—ankylosing spondylitis tuberculosis new growth.

Anterior femoral pain occurs usually in the middle-aged or elderly the probable cause is either arthritis of the hip joint, or femoral neuralgia. X ray films of the hips and lumbar spine are therefore essential If these are quite normal, the condition may of course be a disc protrusion but experience has taught me to repeat the films in 3 or 4 weeks. Some secondary deposits in bone show no radiological change for a few weeks [p. 93]

Meralgia paraesthetica

This is neuralgia and paraesthesia on the outer side of the thigh, generally considered to be due to neuritis of the lateral cutaneous nerve of the thigh. This sensory nerve emerges from the pelvis below Poupart s ligament just medial to the anterior superior iliac spine, and divides into numerous branches supplying the skin on the outer side of the thigh. The condition is rare.

Multiple peripheral neuritis

This is one of the commonest causes of pain in both legs below the knee. Alcoholic neuritis begins with tingling and numbness in the feet, aching legs, night cramps in the calves The legs become weaker with increasing foot drop Diagnosis is discussed on pp 14 15

Lightning pains of tabes dorsalis

Lightning pains are readily recognizable if the patient gives a clear description. The complaint is of a succession of momentary pains, at intervals of a few seconds or minutes, and continued for several hours. They are most often felt in the lower limbs, as if striking the limb at right angles to its surface but they may occur in the trunk and upper limbs. In any one bout the pains are felt in the same region but this varies from day to day The intensity of the pain is not of diagnostic importance

These paroxysms are commonly the earliest symptom and are commonly thought by the patient to be rheumatism especially as they appear to be aggravated by a humid atmosphere. Tabetic patients often complain also of continual dull aching pain in the legs.

Tabes dorsalis affects men much more frequently than women. The commonest age of onset is between 35 and 50 years—usually 5-10 years after the original syphilitic infection

Pain—either typical lightning pains, or a girdle pain in chest or abdomen—is an early symptom The principal physical signs that may be expected early are

1 *Sensory changes* Loss of vibration sense and sense of deep pain in the legs—evoked by firmly compressing the calf muscles or tendo Achillis—are among the earliest. Impairment of sense of position and passive movement comes later

2. *Loss of ankle jerks* and later of knee jerks.

3 *Pupil abnormalities* inequality is common—so is some degree of contraction of the pupils the reaction to light is impaired or lost, that to accommodation normal (Argyll Robertson pupil).

4 *Romberg's sign* This may be obtained before ataxy—usually a late symptom—is recognized The patient stands with heels together eyes first open then closed If he is steady with eyes open, he becomes unsteady when he closes them. A minor degree of Rombergism has been described he remains steady with eyes closed but the extensor tendons on the dorsum of the foot show irregular restless movements (*danse des tendons*) A marked degree of Rombergism is an unsteadiness when standing with eyes open and increased with eyes closed.

This is perhaps the most common syndrome of early tabes lightning pains, loss of ankle jerks loss of vibration and deep pain sense in the legs, Argyll Robertson pupil. Any one of the following, however may be the presenting symptom, or may accompany lightning pains, giving a further clue to the correct diagnosis

1 *Paraesthesia* in the lower limbs numbness in the feet and a sense of 'walking on cotton wool

2. *Failing vision from optic atrophy*

3 *Visceral crises* These are attacks, lasting from a few hours to several days due to excessive visceral contractions vomiting and epigastric pain (gastric crisis) tenesmus (rectal crisis) pain in the bladder and strangury (vesical crisis)

4 *Bladder disturbances* A raised threshold for afferent impulses which produce a desire to empty the bladder results in over-distension, and often residual urine after emptying the bladder This sooner or later

is followed by pyelocystitis. Early symptoms are difficulty in beginning micturition, diminished frequency incontinence at night.

5 *Charcot's Joint* This is a painless arthropathy affecting usually a knee or a hip sometimes the spine or another joint [p 156]

Usually some combination of these symptoms and signs precedes the appearance of frank ataxy by many months. Ataxy begins in the legs walking is slightly unsteady especially in the dark the patient begins to watch his feet, to walk on a broad base with a high-stepping stamping gait. Walking may eventually become exceedingly difficult, or impossible. Ataxy may also appear in the upper limbs [p 6]

A perforating ulcer is usually one of the later events, though it may be early It is a septic ulcer under the big toe or ball of the foot painless and very chronic. It may go very deep

The Wassermann reaction in the blood is positive in 70 per cent. of cases in either blood or C.S F or both in 80 per cent. Characters of the C.S.F of diagnostic value are a moderate excess of mononuclear cells an excess of globulin a colloidal gold curve of the luetic type.

PERIPHERAL VASCULAR DISORDERS

Intermittent claudication [p 142]

This is a symptom so characteristic that its origin in a diminished blood supply to the muscles is at once apparent. But arterial disease may cause pain in the leg which is continual and not related to activity What should direct our attention to the possibility of arterial disease being the cause of pain in the leg are the coincident symptoms and signs of ischaemia—coldness, numbness, colour changes in the skin and in all doubtful cases it should be a routine to examine the arterial pulsations, and postural colour changes [p 144]

Severe, continuous, diffuse and cramp-like pain in the foot and leg is the commonest initial symptom of sudden arterial occlusion [p 43]. This should be thought of when a leg suddenly becomes cold and numb and stricken with severe pain and the femoral and popliteal pulses examined.

Occlusive arterial disease

Arteriosclerosis obliterans and thrombo-angiitis obliterans, if of a degree sufficient to cause severe ischaemia, may cause rest pain. This may be a dull continual aching pain in the foot and lower part of the leg, common at night and badly disturbing sleep. Or it may be the hyper sensibility of the skin of the foot to friction and warmth which also disturbs sleep by burning paraesthesia [pp 45 114] Much more severe are the paroxysmal lancinating pains of ischaemic neuritis, which may be a sequel to sudden arterial occlusion

Arterial and arteriolar constriction

The disorders of peripheral circulation due to constriction of small arteries and arterioles are not as a rule conspicuously painful. In *acrocyanosis* [p 47] the complaint is usually no more than of numbness and coldness In *livedo reticularis* [p 44] there is at most a dull aching of legs and feet. The superficial ulcers of *chronic pernio* may be very painful There may be quite bad pain in severe forms of *Raynaud's disease* [p 45] especially when associated with trophic changes.

Thrombophlebitis [p 71]

Thrombophlebitis of superficial veins in the leg causes a red, tender and painful linear swelling in the skin in the course of a vein. These lesions are often multiple, and tend to appear in crops.

Post-operative thrombophlebitis of the deep veins of the calf is common, and causes pain in the calf and tender calf muscles

Thrombophlebitis of the popliteal vein causes pain and tenderness in the course of the vein, with slight oedema of the foot and lower part of the leg.

Ilio-femoral thrombophlebitis has a sudden febrile onset with quite severe pain in the whole limb there is extensive enlargement of the limb and a widespread pitting oedema. The superficial veins are distended and the skin cyanosed.

Repeated attacks of thrombophlebitis may leave a *chronic venous insufficiency* in the limb which may be a cause of considerable aching.

DISEASES OF BONE

Diseases of bone are discussed in other sections, and are merely summarized here

Tumours [pp 21 7]

1 *Osteogenic sarcoma* At the end of a long bone, most often near the knee. Increasingly severe continual pain, disturbing sleep then the appearance of a swelling.

2. *Ewing's tumour* Patients 5-15 years. Pain and pyrexia intermittently for some months then swelling. Apt to be confused with chronic osteomyelitis.

3 *Giant cell tumour (osteo*

knee. Locally malignant. S

4 *Osteoid osteoma* Be

adults. Localized persistent

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Syphilitic periostitis and osteitis

Continual pain in the legs, of some severity apparently with no physical signs to indicate its probable cause may be a baffling problem until the tibial shafts are examined radiologically A rounded and moderately tender swelling will be at once apparent clinically if the



FIG 44
Syphilitic periostitis
(by kind permission of Dr Cecil Bull).

periostitis affects the subcutaneous surface but the radiological appearance of syphilitic periostitis and osteitis—with dense sclerosis of the affected area—may be found in a part of the shaft that cannot be palpated It is well to have radiographs of the shaft of the bones in all cases of pain in the legs for which no adequate explanation can be found [FIG 44]

OSTEOMYELITIS [p. 157]

GENERAL RAREFYING DISEASES OF BONE [p. 31]

PAGET'S DISEASE [p 28]

MISCELLANEOUS

Referred somatic pain. Aching and pain in the lower limb—particularly the trochanteric area and groin and outer side of the thigh—may be referred from a lesion in the lumbar region. It differs qualitatively from a neuralgia, having none of the paroxysmal quality nor the associated paraesthesiae.

Aching pain, or heaviness in the legs and cramps in the feet, may be an early symptom of *paralysis agitans* [p 61]

Aching of one or both legs is often the earliest complaint in *disseminated sclerosis* [p 213] It is an aching due to attempted normal use of weak muscles. The patient may describe it as tiredness or heaviness of the legs, and may complain also of numbness and tingling.

Subacute rheumatism [p 195]

Subacute rheumatism in children and young adults causes transient and vagrant pains in limbs and joints recurring at intervals, with or without carditis. Growing pains are pains in the lower limbs, often behind the knees where the hamstring tendons will be found to be tender These pains are sometimes severe enough to make the child limp They are troublesome daily for a week or more, then there is a variable interval of freedom before a recurrence. They are especially bad in cold damp rainy weather

Growing pains might be due to chronic fatigue, malnutrition, and postural strain but even if there are no symptoms and signs referable to the heart, it is wise to make a provisional diagnosis of subacute rheumatism—especially if the blood picture shows a hypochromic anaemia, and the E.S R. is raised.

When fleeting pains of this nature are complained of by an adolescent or young adult, the pre-spondylitic stage of ankylosing spondylitis should be considered X ray examination of the sacro-iliac joints will settle the issue [p 87]

Scurvy

Adult scurvy is now very rare, but is seen occasionally when diet has for a long period been grossly deficient in vitamin C. The initial symptoms are lassitude and loss of weight. Then follow characteristic symptoms and signs, mainly due to haemorrhages aching pain in the calf muscles haemorrhages into the muscles, mainly of the legs, causing tender hard swellings petechiae and ecchymoses in the skin congested and bleeding gums The platelet count is not reduced. Administration of ascorbic acid, 150-300 mg. daily is a valuable therapeutic test.

Infantile scurvy appears usually between the ages of 6 and 18 months, in infants having artificial feeding deficient in vitamin C The earliest symptom, sudden in onset, is severe pain in the legs from subperiosteal haemorrhage In addition to the haemorrhagic tendency there is defective bone formation [p. 37]

The anterior tibial syndrome

This is a severe pain in the pre-tibial region coming on during strenuous activity and due to ischaemic necrosis of the anterior tibial and extensor muscles Reported cases[1] have been mainly in healthy young soldiers during a route march or in young men playing football. The sequence of events is pain in the front of the leg—swelling of the anterior tibial compartment and reddening of the overlying skin—inability to dorsiflex the foot and toes from paralysis of the tibialis anterior and extensors of the toes. Muscle biopsy has shown a massive ischaemic necrosis.

It is well known that pre-tibial pain is common in athletes beginning training, and it is thought that muscle strain initiates the anterior tibial syndrome being followed by tension in the rigid anterior tibial compartment sufficient to impede the circulation, and lead to ischaemic necrosis.

PAIN IN THE SHOULDER

The shoulder in common speech means the whole area of the shoulder girdle, and often the side of the neck as well Find out first where pain is felt most intensely whether cervico-scapular or deltoid. Then the character of the pain will be clarified and thus, with its duration and associated symptoms may take one half way to a diagnosis.

The clinical examination is designed to locate the operative lesion for a small lesion may give widespread pain. The minimum data required are 1 the range of movement in the cervical spine 2. the condition of the shoulder girdle muscles 3 the range of passive movements at the scapulo-humeral joint. Active movements are not of much diagnostic value Passive movements are tested with the patient recumbent, so that the scapula is fixed on the couch by the weight of his body

This is usually enough for a tentative diagnosis of the nature of the lesion. But it will in all cases be confirmed, or otherwise, by the positive or negative data provided by X ray films of the cervical spine and shoulder joint, or by other clinical or radiological studies mentioned below in the appropriate context

[1] Barham, C. A., *et al* (1949) *Lancet* ii, 928

CERVICO-SCAPULAR PAIN

The main causes are

OF SOMATIC ORIGIN

In the cervical spine—referred pain or root pain

cervical disc protrusion

cervical spondylosis

disease of the cervical vertebrae

tuberculosis osteomyelitis new growth ankylosing spondylitis

acute brachial radiculitis

herpes zoster and post herpetic neuralgia

In the shoulder girdle muscles

fibrositis

paralysis agitans

hemiplegia

winged scapula [p 20]

paresis of the deltoid

OF VISCERAL ORIGIN

apical or diaphragmatic pleural pain

involvement of T 1 nerve root in apical carcinoma

It is scarcely possible to consider cervico-scapular pain without reference to disorders in which pain is felt chiefly in the neck and these are included here.

PAIN ORIGINATING IN THE CERVICAL SPINE

Cervical disc protrusion

This is one cause of painful stiff neck, which may or may not be followed by brachial neuralgia [p. 74]

Cervical spondylosis

Cervical spondylosis is the name generally used for a condition of the cervical spine due primarily to disc degeneration. This is readily seen in X ray films, being indicated by narrowing of the disc space and the secondary bone reaction, including lipping of the adjacent vertebral bodies [FIG 45] Oblique films may show osteophytes invading the foramen. At the level of the degenerated discs and possibly at other levels a fibrotic thickening has been described of the dural root sleeve sheathing the nerve root. This constricting ring round the nerve root, and at times narrowing of the foramen by an osteophyte, is held to be responsible for nerve root syndromes.

Cervical spondylosis is thought to be responsible for the following syndromes

Root pain and referred pain. It may be difficult to distinguish the two for the pain of a sensory nerve root irritation and referred pain from the lesion without root involvement are both felt *in the same somatome.* The two pains differ in character neuralgia is a lancinating pain in



FIG. 45
Cervical spondylosis disc degeneration C.3-4 and C.5-6-7

paroxysms, especially bad at night. But with an unreliable witness this is a poor guide more helpful is a complaint of paraesthesia—tingling and numbness—in the hand, for this is an almost constant accompaniment of neuralgia, and *is absent in referred pain.*

The pain, whether radicular or referred, is deep pain and less noticeably skin pain. To establish that the area of pain is in such and such

dermatomes is therefore of no help in locating the operative lesion. The cervico-scapular area represents roughly the dermatomes C.3 4 T 1 2 and 3 The intervening dermatomes and part of T 1 and 2, are found in the arm. It might therefore be concluded that pain felt solely in the posterior part of the neck and shoulder and not at all in the upper limb must arise at the levels C.3 4 or T.3 and that root pain from C.5 to T.2 would be felt solely in the arm But this is not so It applies to paraesthesia, but not to pain to note the dermatomes affected by paraesthesia is at times of diagnostic value

The somatome of a nerve root includes the muscles, fasciae, periosteum, joints, ligaments, and so on, to which its sensory fibres are distributed and these cover a much wider area than the dermatomes.

In cervical spondylosis the discs most commonly affected are C.5-6 and C.6-7 the corresponding nerve roots being C.6 and C.7 The pain from either lesion is felt in the back of the neck and cervico-dorsal region, the scapular and often the pectoral regions, as well as, in root pain and in very severe referred pain, in the arm.

Acroparaesthesia [pp 186-8].

Muscular wasting from anterior root pressure, which when painless may suggest motor neurone disease

Subacromial bursitis This is possible but not proven. The two are often found in association, but a causal association is not established.

Myelopathy [p 214] Symptoms may arise from direct compression of the cord or from a local ischaemia in the cord due to compression of the anterior spinal artery symptoms that may mimic motor neurone disease, disseminated sclerosis, subacute combined degeneration, and extra and intra medullary tumours. For a long time it may cause only paraplegia, with no abnormality in the upper limbs.

Disease of the Cervical Vertebrae Even if accompanied by root pain, tuberculosis, osteomyelitis, and new growth in the bone of the cervical spine at once direct attention to the neck rather than the shoulder by the severe local pain and restricted movement [pp 77 78]

In *ankylosing spondylitis* the same observation applies The cervical part of the spine may be affected first but more often it is a late event.

Acute brachial radiculitis

This disorder occurs in minor epidemics, and is probably a virus infection. The first symptom is pain, usually severe, in the root of the neck and shoulder After several days, when pain is abating, weakness and wasting of some of the shoulder girdle muscles develop There is usually an area of sensory loss over the deltoid. Recovery of power is

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slow Eventually after a year or two very marked improvement comes in most cases though in many some atrophy remains. The serratus anterior may be involved, giving a winged scapula [p 20], and this muscle is particularly slow to recover

Serum Radiculitis or Neuritis This has been observed many times as a complication of prophylactic serum injections, especially anti tetanus serum.[1] It affects especially the lower cervical roots the symptoms are as described above. Muscular weakness and atrophy affect chiefly the deltoid spinati, serratus anterior biceps, brachialis, and triceps. Rarely the deltoid, or the serratus, alone is affected. That the condition is an allergic phenomenon is suggested by the prompt relief brought by administration of adrenal steroids.[2]

PAIN ORIGINATING IN THE SHOULDER GIRDLE MUSCLES

What we understand by *fibrositis* is discussed on p 3 It probably accounts for a large number of instances of shoulder girdle pain. But it has so few distinctive physical signs that it should not be diagnosed until all other possibilities have been excluded.

The muscular rigidity of *paralysis agitans* [p 61] or of *hemiplegia* [p 210] is often associated with a good deal of general aching or even pain in the shoulder girdle A complaint of aching after use of the arm when there is *paresis of the shoulder girdle muscles* is to be expected.

PLEURAL PAIN

Prolonged aching in the cervico-scapular region in which none of the causes mentioned can be found or cases taken to be fibrositis which show no response to treatment, should suggest the possibility of chest disease. This applies particularly to dorsal pain.

Pleural pain is felt over the area of pleura involved except when this is the dome of the diaphragm. *Apical pleural pain* may be caused by apical tuberculous, and is felt as a dull persistent ache in that situation. The dome of the diaphragm is supplied by the phrenic nerve derived from C.3 4 5—mainly C.4 so that *diaphragmatic pleural pain* may be referred to the cervico-scapular region.

It is therefore obvious that when persistent pain in the shoulder and especially in any part of the chest wall, is unaccountable, X ray examination of the chest is essential.

[1] Miller H. G., and Stanton, J B. (1954) *Quart J Med.*, 23, 1
[2] Baron, J H. (1958) *Brit med. J.*, ii, 678.

PAIN IN THE DELTOID REGION

By this we mean pain felt chiefly in the deltoid region, even though it is felt also in the cervico-scapular region and down the arm.

The main causes are

SOMATIC ORIGIN

The rotator cuff and subacromial bursa

- supraspinatus tendinitis
- calcification in the supraspinatus tendon
- chronic subacromial bursitis
- acute subacromial bursitis
- the shoulder hand syndrome

Joints

- scapulo-humeral joint rheumatoid arthritis osteo-arthritis tuberculous arthritis ankylosing spondylitis
- acromio-clavicular joint osteo-arthritis

Bone

- osteogenic sarcoma of the upper end of the humerus

REFERRED VISCERAL PAIN

Left shoulder angina

Right shoulder subphrenic inflammation

THE ROTATOR CUFF AND SUBACROMIAL BURSA

The flattened tendons of the four short rotator muscles blend with the capsule of the shoulder joint before their insertion into the humerus they are, anteriorly the *subscapularis* superiorly the *supraspinatus* and posteriorly the *infraspinatus* and *teres minor* They form a musculo-tendinous cuff to the joint.

The *subacromial bursa* forms an auxiliary shoulder joint between the *upper end of the humerus and the arch of the acromion process* and coraco-acromial ligament [FIG 46] It is liable to become inflamed in lesions of the musculo-tendinous cuff which forms a large part of its floor Subacromial bursitis mimics arthritis of the scapulo-humeral joint in that passive movements in every direction are restricted. When disease of the bone and joint can be excluded this sign is diagnostic of bursitis, for it is not found in uncomplicated lesions of the musculo-tendinous cuff.

The action of the short rotator muscles is synergic they hold the head of the humerus in the glenoid during the action of the prime mover which effects rotation or abduction. They all act in some degree in this way in all movements of the arm at the shoulder joint.

Supraspinatus tendinitis

This is the commonest lesion of the rotator cuff and probably the commonest of all minor disabilities of the shoulder Its pathology is obscure, though many instances are undoubtedly traumatic. It is probable, however that sprains and partial ruptures are preceded by

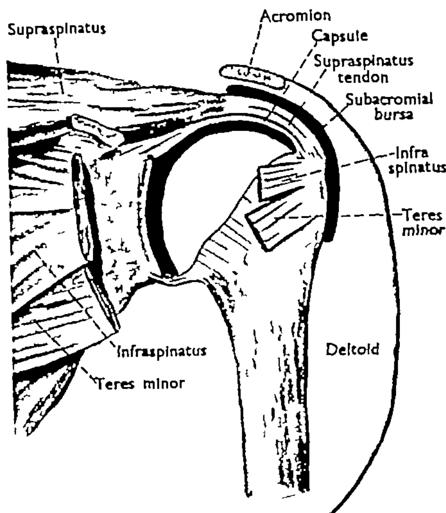


FIG. 46
Drawing of subacromial bursa showing its relation to the supraspinatus tendon and the shoulder joint.

degenerative changes in the tendon. Sleep disturbed by intolerable aching in the shoulder when lying on it is as frequent and prominent a symptom as the more expected one of a catch of pain in the shoulder when putting on a coat, or performing comparable movements. Soon

every use of the arm brings a wince of pain and active movements become restricted the hand cannot be carried behind the back with its usual facility nor can it reach up as high as the sound limb

A tentative diagnosis can be made on these symptoms. It will be confirmed by these observations

1 Active abduction is accompanied by a small painful arc of movement at about 90° abduction

2 There is a tender point between the tip of the acromion and the humeral tuberosity

3 Passive abduction and external rotation are either slightly restricted or of full range but in either case sharp pain is felt at the limit of movement.

4 X ray films of the shoulder are either normal, or show no more than a zone of decalcification in the greater tuberosity of the humerus

Calcification in the supraspinatus and short rotator tendons

A focal zone of degeneration in the musculo-tendinous cuff may calcify Calcareous deposits are usually single, and usually in the supra spinatus tendon close to its insertion. But there may be two or more, and they may form in the tendon of the subscapularis, infraspinatus or teres minor

The symptoms may be very much those of supraspinatus tendinitis. But pain is usually more severe and may be more constant. It is felt most intensely in the region of the insertion of the deltoid The severity of the symptoms and signs—as with simple tendinitis—depends on the degree of associated inflammation of the subacromial bursa. Symptoms may last for many months, the deposit being eventually absorbed, or ceasing to cause symptoms At times spontaneous cure is brought about by a *fulminating seizure of acute bursitis. The calcified zone becomes* acutely inflamed calcareous material softens, escapes into the bursa and precipitates an acute bursitis. Absorption from the bursa then occurs in a few weeks.

Palpation may reveal a tender point near the insertion of the supra spinatus or in supscapularis deposits there may be a point of tenderness medial to the bicipital groove. Passive movements at the scapulo-humeral joint are restricted to a degree varying with that of the associated bursitis they may be severely restricted in all directions if intra-bursal adhesions have formed.

The deposit appears in X ray films as an abnormal shadow usually in the region of the supraspinatus tendon (FIG 47] Many subscapularis and infraspinatus deposits may not be seen because the shadow and that of the humeral head overlap

Acute subacromial bursitis (FROZEN SHOULDER)

This may complicate calcification of the musculo-tendinous cuff or may occur as a primary disorder It affects men and women equally and is not often seen at a younger age than 40 The shoulder is gripped by agonizing pain, and becomes immobile from pain and spasm. The patient hugs his arm to the side and flinches from any attempt by the examiner to move it The whole area of the subdeltoid part of the bursa is acutely tender There is a fibrinous exudate in the bursa, but nearly



FIG. 47

Calcareous deposits in the subacromial bursa—and probably in the supraspinatus tendon.

always so little than it cannot be detected clinically With rest, acute pain abates in about 2 weeks and is followed as a rule by a long period of disability from chronic bursitis

The signs and symptoms are those of an acute arthritis of the shoulder But monarticular arthritis of the shoulder especially acute arthritis, is very uncommon. It will usually show suggestive X ray appearances whereas X ray films in primary acute bursitis show no abnormality other than some decalcification of the greater tuberosity

The shoulder-hand syndrome [p 84]

This begins with much the same symptoms as those of acute subacromial bursitis. It is recognized by the subsequent pain swelling and stiffness of the hand and fingers, and the trophic changes.

The syndrome of Forestier and Certonciny

Under the name *pseudo-polyarthrite rhizomélique* Forestier and Certonciny of Aix-les-Bains[1] have described an important syndrome, which is undoubtedly a distinct clinical entity Perhaps it would be better named rhizomelic peri-arthritis. It is a disorder of the second half of life, affecting both men and women. Constitutional disturbance and pyrexia are common the temperature may rise to 100-101° F It is characterized by severe pain and restricted movement in both shoulders, and in less degree in the hip and lumbar regions. No other joints are affected apart from fleeting joint pains.

There are no significant radiological changes in shoulder hip or sacro-iliac joints. The E.S R. is always increased usually considerably sometimes in the region of 100 mm. in one hour There may be a moderate anaemia, and rarely a leucocytosis. Although not progressive, spontaneous recovery may be exceedingly slow Improvement under adrenal corticosteroids is startlingly prompt pyrexia abates, pain stops, the patient quickly feels well, and normal mobility of the shoulders is gradually restored. Relapse will probably follow if steroid treatment is stopped in less than 6-9 months. Gold treatment is said to be effective.

The syndrome can be distinguished from a rhizomelic form of rheumatoid arthritis. No synovial swelling or effusion can be detected in the shoulders, all patients recover with no residual joint signs, and recurrence is unknown. It is also clearly not the common subacromial bursitis, nor ankylosing spondylitis, nor one of the known collagen diseases

[1] Forestier J., and Certonciny A. (1953) *Sem. Hop Paris*, 34, 3220.

JOINTS

The scapulo-humeral joint

Osteo-arthritis of the scapulo-humeral joint is almost a rarity It is found mainly in polyarticular osteo-arthritis, especially in the elderly It may be secondary to a congenital malformation of the humeral head or severe trauma. The clinical features—dull pain in the shoulder atrophy of the deltoid and spinati and restricted movement—are similar to those of chronic subacromial bursitis, to the slowly progres-



FIG. 48
Osteo-arthritis of the shoulder

sive shoulder stiffness of senile tuberculosis of the joint, to rheumatoid arthritis, and the arthritis of ankylosing spondylitis. A radiograph shows characteristic changes [FIG 48]

The only true arthritis affecting the shoulder joint alone is *tuberculous arthritis* The patient is usually young—but it may occur in the elderly —and the complaint is of gradually increasing stiffness of the shoulder For some time there is little or no pain. Demonstrable synovial effusion is absent and peri articular abscess is rare. A suggestive diagnostic feature is very marked wasting of the shoulder girdle muscles. X ray films show quite early extreme decalcification of the humeral head Later there is erosion of the articular surface progressing to considerable destruction [FIG 49].

Arthritis of the shoulder may occur as part of a *rheumatoid polyarthritis* [FIG 50] One joint only may be affected first in rheumatoid arthritis, but this is scarcely ever the shoulder joint.



FIG. 49
Tuberculosis of the scapulo-humeral joint in a man aged 33

The shoulder may also be affected by a rheumatoid type of arthritis as a late event in *ankylosing spondylitis*

The acromio-clavicular joint

The acromio-clavicular joint is functionally of little importance. It is liable to osteo-arthritic changes, which may at times be responsible for a mild shoulder pain on movements of the limb Other causes of pain should be excluded, for the condition is often symptomless.



FIG 50

Rheumatoid arthritis of the scapulo-humeral joint hand and shoulder from the same patient.

BONE

Osteogenic sarcoma of the upper end of the humerus

This affects children, adolescents and young adults most instances occur in the second decade. The upper end of the humerus is a common site—the second commonest after the region of the knee The mere fact of an adolescent complaining of severe pain in the deltoid region is disturbing adolescents do not get acute subacromial bursitis tuberculosis of the shoulder is not very painful and any of the serious diseases of the cervical spine would give root pain over a much wider area. If pain is soon followed by a palpable swelling of elastic consistency the most careful and expert radiological studies are essential.

If such time has passed that the X ray appearances are easily recognized, so probably has opportunity for successful treatment. In the early stages, even though there is a palpable tumour X ray changes may be so slight that great experience is needed for diagnosis. Moreover radiological appearances vary considerably with different types of growth. Characteristically there will be seen under the soft tissue swelling a little mound of periosteal new bone where the periosteum has been raised from the shaft Its central part is eroded. Spicules of periosteal new bone may be seen at right angles to the shaft. It is well known that sarcoma of bone may follow trauma and these appearances might represent an ossifying haematoma with local periostitis. We might also be dealing with an abscess, or early osteomyelitis.

The shaft in the affected area shows some increase in density and a loss of detail of the cancellous bone structure. This can be seen by comparison with the bone of the epiphysis, which for some time escapes. An *osteoplastic* type of growth—in which bone formation exceeds bone destruction—gives a dense sclerosis of the affected bone. With some *osteolytic* growths there is no new bone formation, but merely an erosion of the cortex and destruction of underlying bone, under the soft tissue tumour In all suspicious cases the lungs are X rayed for possible metastases

Diagnosis based on clinical and radiological data should always be tested by biopsy

REFERRED VISCERAL PAIN

Angina

Angina is very unlikely to be a source of confusion in the diagnosis of a painful shoulder The type case is a patient over 40 who gets recurrent attacks of a dull heavy ache and sense of pressure behind the sternum, and pain which may spread to the left shoulder and down the

arm, and to the left side of the neck. Attacks are usually brought on by exertion, or by emotional disturbance, and do not last longer than a few minutes.

Subphrenic Inflammation

A belief persists that *shoulder tip pain is a symptom of gallstones* and chronic cholecystitis a belief reinforced by the knowledge that the phrenic nerve (from C.3 4 5) sends a few twigs to the gallbladder But this is an error The only shoulder pain associated with gallstones and chronic cholecystitis is that felt at the inferior angle of the right scapula and in the right interscapular region

Pain in the region of the right acromion does occur in inflammatory conditions on the under surface of the right diaphragm, as in subphrenic abscess, or spread of infection from acute cholecystitis but it is a most unimportant feature of a serious symptom complex. Sensory fibres of the phrenic nerve are distributed to the diaphragmatic pleura. Their stimulation causes pain in the region of the acromion, supplied by the supraclavicular nerve from C.3 and 4

Gallstones, or chronic cholecystitis, is not a possible diagnosis for an obscure deltoid pain.

PARAESTHESIA

Paraesthesia means an abnormal sensation and relates to either pain touch or temperature sense. Modifying pain sensation it is the coarse tingling called in popular speech 'pins and needles Numbness is the corresponding abnormality of touch. When what would normally be felt as warmth is felt as a painful burning sensation, we have the corresponding perversion of temperature sense.

Paraesthesia indicates some lesion of the sensory pathways, *usually* the peripheral nerves. The lesion may be ischaemic, which *explains* the common occurrence of paraesthesia in peripheral vascular disease

The common causes of paraesthesia are

PERIPHERAL VASCULAR DISEASE

- Arteriosclerosis obliterans [p 144]
- Thrombo-angiitis obliterans [p 145]
- Arterial occlusion [pp 43, 73]
- Erythromelalgia [p 114]

NERVE ROOT AND PERIPHERAL NERVE LESIONS

Upper limbs

Brachial neuralgia [p 74]

Cervical rib syndrome [p 81]

Acroparaesthesia cervical spondylosis compression of the median nerve in the carpal tunnel costo-clavicular syndrome

Ulnar neuritis [p 12]

Lower limbs

Sciatic neuralgia [p 161]

Upper and lower limbs

Multiple peripheral neuritis [p 14]

CENTRAL NERVOUS SYSTEM LESIONS

Subacute combined degeneration

Disseminated sclerosis [p 213]

Tabes [p 165]

TETANY [p. 49]

In most of these conditions paraesthesia is not the presenting symptom for which the patient seeks advice this is much more likely to be pain or muscular weakness and atrophy In one of them it is the dominant symptom acroparaesthesia. It is a prominent symptom in erythromelalgia, multiple peripheral neuritis, and early subacute combined degeneration.

ACROPARAESTHESIA

Acroparaesthesia names a common syndrome affecting mainly middle-aged women the complaint is of nocturnal attacks of painful tingling or numbness in some or all of the fingers. It is now accepted that compression of the median nerve in the carpal tunnel is its usual cause. Some say it is the only cause but the evidence is not entirely convincing. There are grounds for believing that either cervical spondylosis, or costo-clavicular compression is responsible in some instances.

Cervical spondylosis [p 172] is frequently found in association with acroparaesthesia, and appropriate treatment is often followed by relief Neither fact, however provides very strong evidence of causal relation-

The costo-clavicular syndrome

Fatigue and loss of tone in the muscles elevating the shoulder girdle will subject the brachial plexus and subclavian artery to intermittent pressure between the clavicle and the first rib During the Second World War it was a fairly common observation that a recruit of poor physique

would complain of pain and numbness and congestion in his arms and hands when carrying an army pack Recovery was prompt after removing the weight from his shoulders and after his shoulder girdle muscles had been strengthened by suitable physical training he was no longer subject to this disability It was another common observation of the war years that women doing their shopping and housework for the first time were apt to complain of numbness, tingling and uselessness of the fingers

Such compression is, of course, especially apt to occur when the space between the clavicle and the first rib is unusually narrow it is indeed doubted that compression can occur if anatomical conditions are normal Some attacks of acroparaesthesia are it is suggested the effect of costo-clavicular compression from fatigue of the shoulder girdle muscles, seen most often in middle-aged women who continue to do heavy work in a state of chronic fatigue Rest, and exercises to improve the tone of these muscles, are often followed by relief

Compression of the median nerve in the carpal tunnel

This is the most probable diagnosis, it is now believed if 1 attacks of painful paraesthesia are nocturnal, and there is little or no discomfort during the day 2. paraesthesia affects two or more of the first four digits, but not the little finger

The right hand is usually the first affected in most instances the disorder becomes bilateral later Patients relate that they wake almost every night, sometimes two or three times a night, with tingling or numbness in the fingers, accompanied by pain which may spread to the palm and forearm They must get out of bed and swing the arms, to get relief which comes in about 10-30 minutes These nocturnal attacks often become more frequent and more severe as time goes on

A few patients show wasting of the thenar muscles. In about one half there is some blunting of skin sensation within the median distribution in the hand.

Compression of the median nerve in the carpal tunnel was first described in 1947 [1] A report on 40 cases submitted to operation was published in 1953 [2] Its conclusions find support in later publications [3] that complete relief in practically all cases follows division of the transverse carpal ligament. The nerve at operation has often been seen to be compressed beneath the ligament, and swollen for a short distance above it.

It seems fairly clear that the diagnosis is justified if and only if pain and paraesthesia have a strictly median distribution.

[1] Brain, R *et al.* (1947) *Lancet* i, 277
[2] Kremer M., *et al* (1953) *Lancet* ii, 590.
[3] Dick, T B. S and Zadik, F R. (1958) *Brit med. J.*, ii, 288.

What causes compression in the carpal tunnel is obscure. In a recently published series of 35 consecutive patients with myxoedema,[1] 26 complained of acroparaesthesia. Symptoms and signs had a strictly median distribution and usually cleared up with thyroid treatment. The suggestion is that the syndrome can be caused by accumulation of myxoedematous tissue under the transverse carpal ligament.

MULTIPLE PERIPHERAL NEURITIS AND SUBACUTE COMBINED DEGENERATION

One of the earliest complaints in alcoholic neuritis is of tingling and numbness in the feet and toes So it is in most other forms of multiple peripheral neuritis [p 14]

Subacute combined degeneration is a condition of degeneration of the posterior and lateral columns of the cord, and of peripheral nerves. It is associated with pernicious anaemia in the middle-aged and elderly There are two types the flaccid type in which peripheral nerve changes predominate the spastic type in which there is more severe involvement of the cord

The *flaccid type* is the commoner and may at the onset be indistinguishable from multiple peripheral neuritis The complaint is of tingling and numbness in the feet, and to a less degree in the hands, with weakness and ready fatigue of the legs. On examination, knee and ankle jerks are not obtained In all cases the calf and plantar muscles are tender Sensory ataxy soon appears, in an unsteady gait and clumsiness of the fingers This picture in the middle aged and elderly calls for a complete blood count, and possibly gastric analysis The clinical symptoms and signs of pernicious anaemia are often obvious but in some instances neurological signs appear first, even before the blood changes. An achlorhydria is an almost constant finding.

The diagnosis may depend on evidence of cord involvement a severe loss of postural sensibility and vibration sense, and the appearance of an extensor plantar response Tender calf muscles with signs of spinal cord disease are almost pathognomonic.

The *spastic type* is referred to on p 215

PARAESTHESIA AND PERIPHERAL VASCULAR DISEASES

The first symptom of *sudden arterial occlusion* in a limb [p 43] is not always an attack of extremely severe pain the earliest indication may be numbness and tingling. Not only should we examine arterial

[1] Murray I P C and Simpson, J A. (1958) *Lancet*, i, 1360.

pulsation as a routine procedure in all cases of sudden excruciating pain in a limb but also in numbness and tingling of sudden onset especially when associated with coldness and pallor of the extremity

Symptoms from *arteriosclerosis obliterans* [p 144] are nearly always in the lower limbs usually for many months in one only Early symptoms include, besides coldness of the limb and weakness numbness and tingling in the foot, and perhaps burning paraesthesia.

Thrombo-angiitis obliterans [p. 145] occurs almost exclusively in men, between the ages of 25 and 40 Coldness and numbness of a hand or foot is a common early complaint there may also be tingling and burning paraesthesia

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POLYARTHRITIS

CHRONIC POLYARTHRITIS

Chronic arthritis affecting two or more joints is likely to be either osteo-arthritis or rheumatoid arthritis. There are many possibilities to be considered in the differential diagnosis of rheumatoid arthritis recognition of polyarticular osteo-arthritis is relatively simple.

POLYARTICULAR OSTEO ARTHRITIS

This is a generalized ageing process in joints varying in degree from joint to joint, and in some sufficiently marked to give symptoms.

It affects men and women but the type case is a woman aged about 50 probably complaining of pain everywhere The hands are painful, and many of the finger joints are now enlarged and have lost their suppleness there are, or have been, episodes of pain in the shoulders and back (referred from the apophyseal joints of the spine, often affected) and there may be complaint of pain in feet and knees

Serious crippling is rare, and the general health is not affected, as it is in rheumatoid arthritis.

The hands differ from those of rheumatoid arthritis the diagnosis can often be made solely from their appearance [p 123] The knees are commonly affected and in severe cases hips, elbows, shoulders and spine

Radiology The X ray appearances are quite characteristic but it is important to know that old rheumatoid joints may show secondary osteo-arthritic changes. There will be found 1 diminution of joint space, usually asymmetrical 2. increased density of the articular lamella 3 in the subarticular bone, irregular sclerosis side by side with

local osteoporosis and perhaps cavity formation 4 osteophytes [FIG 51]

Laboratory Findings The E.S R is usually normal, or only very moderately increased. There are no changes in the blood picture, or blood chemistry



FIG. 51
Osteo-arthritis in the hand.

RHEUMATOID ARTHRITIS

This is a systemic disease—one of the collagen diseases—whose main impact is on the joints The type case is a young woman, who at the onset complains of transient pains in fingers, wrists, feet or knees and of a feeling of stiffness in these joints at the beginning of the day until use has restored a freer movement. Joint pains may last only a day or two but return after some weeks to stay longer and in time they return with a more lasting stiffness and some joint swelling

The joints of the hands are often the first to show signs of frank arthritis at the same time the feet may be tender and swollen in the metatarso-phalangeal region. The progress is often wrists, tarsal joints, knees, elbows, and shoulders, in this order As new joints become seized, those first affected are more clearly involved in the morbid process. The appearance of the hands may be almost pathognomonic. They are affected symmetrically first the proximal interphalangeal joints, then metacarpo-phalangeal, intercarpal, and wrist joints. The pale, clammy hands, with swollen fingers intercarpal and wrist joints thrown in high relief by muscular wasting, and with ulnar deviation of fingers and wrists from uneven muscular spasm, are very characteristic.

Any typical rheumatoid joint feels soft and doughy in regions where the capsule is palpable usually the amount of synovial effusion is not large, the swelling being more synovial thickening and a gelatinous exudate Sometimes a general enlargement of the bone ends contributes to the joint swelling. The overlying skin is neither red nor hot, for the affection is essentially chronic.

Painless subcutaneous nodules, sometimes mobile but more often fixed to deeper fibrous tissues, may appear over bony prominences during the course of rheumatoid arthritis. They are most often found over the subcutaneous border of the ulna, within an inch or two of the olecranon. Most of these nodules seem to be permanent, or at least to persist for many years—in contrast to those of rheumatic fever

The constitutional nature of rheumatoid arthritis is seen in the almost constant disturbance of general health. This varies considerably in degree. When the disease is active there are symptoms of anaemia, loss of weight, and bouts of intermittent pyrexia, the temperature rising to 99-100° F in the evenings.

Early in the illness spontaneous remission is common, lasting from a few months to several years it occurs in about 25 per cent of untreated cases. But once well established the disease may progress relentlessly

Disease of the Eye in Rheumatoid Arthritis Uveitis is very uncommon in rheumatoid arthritis, in contrast to its comparative frequency in ankylosing spondylitis and in Reiter's disease

In 1933 Sjögren described a rare syndrome—since known as Sjögren's syndrome—of rheumatoid arthritis and keratoconjunctivitis sicca, and at times rhinitis and pharyngitis sicca and gastric anacidity The patient complains of dry sore eyes, dry nose and mouth, and hoarseness. The salivary glands are enlarged

Radiology Early in the disease there may be no demonstrable radiological changes in the affected joints, except soft tissue swelling readily seen in the films. But often there is a general osteoporosis of phalanges,

metacarpals and carpus and thus is very characteristic. In the established disease this rarefaction is constant the 3 zones of varying bone density of a normal phalanx or metacarpal become almost uniform. Early there may be found slight loss of joint space. Not until later do erosions of the articular surface and the marginal areas appear Cystic areas may be seen in the subchondral region [FIG 52] All these changes are progressive.

There are no osteophytic outgrowths, except in the advanced disease where secondary osteo-arthritic changes have occurred.

Laboratory Findings The E.S R. is always raised If it does not show at least a moderate rise, a diagnosis of rheumatoid arthritis is improbable By the Westergren technique the upper limit of normal is 3 mm. in 1 hour in men, and 7 mm in women. But in clinical practice these figures are too low errors will be avoided by regarding an increase to 10 mm. in 1 hour in men, or 15-20 mm in women, as unimportant and probably transient. Only a figure above these should be taken to be a significant increase up to 35 is considered moderate, 35-75 marked, and above 75 very marked

Alteration in the pattern of plasma proteins, determined by the paper electrophoretic method, occurs in rheumatoid arthritis. But so it does in all the collagen diseases and many unrelated conditions it is therefore of little diagnostic value. There is a decrease in albumin, and an increase in the α_1 α_2, and γ-globulins and in fibrinogen. There is thus a reversal of the normal albumin/globulin ratio The increased fibrinogen is considered to be largely responsible for the raised E.S R. Natural remission, and clinical improvement under steroid therapy are associated with return to a more normal protein pattern.

Anaemia of a hypochromic and normocytic type is very common, the degree varying with the activity of the disease.

There is no specific laboratory test for rheumatoid disease but the sheep cell agglutination test (the Waaler Rose test) may be helpful In rheumatoid disease there is in the serum a factor in relatively high titre that agglutinates sensitized sheep cells—i e sheep erythrocytes suspended in a non-agglutinating amount of anti sheep erythrocyte serum.

A recent report[1] gives the following information In definite rheumatoid arthritis the test was positive in 87 per cent. of cases when typical rheumatoid nodules were present it was positive in 100 per cent. of males and in 92 per cent. of females In rheumatoid arthritis of less than one year s duration (when a diagnostic test is most needed) it was positive in 70 per cent. In juvenile rheumatoid arthritis only 40 per cent. were positive. In both systemic lupus erythematosus and systemic

[1] Kellgren, J H *et al* (1959) *Brit. med J.*, i, 523

sclerosis 41 per cent. of cases gave a positive result. In many other conditions—e g. osteo-arthritis tuberculous arthritis, Reiter's disease rheumatic fever and ankylosing spondylitis—positive results occur but the percentage is very low

At times aspiration and examination of synovial effusion may be of



FIG. 52.
Advanced rheumatoid arthritis in the hand bone of normal density shown above.

some help Joint fluid for the laboratory should be collected in three containers 1 a sterile tube for bacteriology 2. an oxalated tube for cytology 3 a plain tube for chemistry

Effusions of trauma and of osteo-arthritis show a total cell count of much below 10 000 per c.mm —often below 1 000 and a differential count shows a preponderance of mononuclears. The exudate of rheumatoid arthritis and of any infective arthritis shows a total count of well above 10,000 per c.mm., and characteristically polymorphs predominate.

Cytology of the effusion may therefore help to distinguish a mon-articular rheumatoid arthritis of the knee from traumatic synovitis it is of no help in distinguishing rheumatoid and tuberculous arthritis.

Biopsy of synovial membrane may show pathognomonic histological features. The tissue is swollen and hyperaemic the swelling mainly due to an abundant granulation tissue though some of it represents an increase in size of the synovial villi, and a new formation of numerous secondary villi. The granulation tissue shows the changes of any chronic inflammatory process—numerous lymphocytes, plasma cells and fibroblasts in a matrix of newly formed connective tissue fibres and numerous capillary vessels—and in addition a number of focal collections of lymphocytes in closely packed clusters.

Biopsy is perhaps most helpful in excluding tuberculous infection.

Differential diagnosis in established rheumatoid arthritis

No difficulty should arise in recognizing typical rheumatoid arthritis. Osteo-arthritis, with which it was once confused, is easily distinguished. It can, sometimes legitimately be confused with

1 *Other causes of deformed hands and trophic changes in the hands* [pp 56 128]. In this group I may mention *sclerodactyly* (acrosclero-derma) In the beginning this disease can very easily be mistaken for rheumatoid arthritis it occurs most often in young women, and very often at the onset is associated with Raynaud s phenomenon. The first complaint is of aching swollen fingers, which have lost their suppleness, and of blanching of the fingers Probably in most instances sclerodactyly is for some time treated as rheumatoid arthritis, but as time goes on, the skin of the fingers, and later the back of the hands and forearms, is seen to be tight and unwrinkled like tightly stretched parchment. Swollen at first, the fingers become thinner from loss of subcutaneous fat and trophic changes such as cracked nails, fissures and small ulcers appear on the finger tips.

There is a flexion deformity of the fingers with radiologically normal joints in time the fingers become very stiff perhaps completely fixed. Sometimes subcutaneous nodules of calcium form on them, occasionally extruding through the skin.

The pathological change in the skin is a loss of the normal structure of connective tissue collagen fibres, which are gradually replaced by a dense fibrosis.

2. *Other types of polyarthritis* The hands of chronic polyarticular gout may superficially resemble those of rheumatoid arthritis but (i) joint swelling is harder and more nodular (ii) there is little muscular wasting and no ulnar deviation (iii) peri-articular tophi may be present

(iv) there is probably a history of acute episodes in the foot (v) plasma uric acid is increased (vi) X ray films may be suggestive [p 126]

There is reason to believe that chronic polyarthritis associated with gonorrhoea, which may resemble rheumatoid arthritis, is, at least in most instances, Reiter s disease [p 207]

Two or more joints may be affected by a typical tuberculous arthritis, but this is rare. If a polyarthritis appears in a patient with a tuberculous lesion, or a history of tuberculosis, a synovial biopsy is probably indicated It is wise to get X ray films of the chest in every case of supposed rheumatoid arthritis.

Diagnosis at the onset of rheumatoid arthritis

The real diagnostic difficulties occur 1 before the appearance of frank arthritis 2. when the onset course, or joint pattern of rheumatoid disease are atypical.

Arthralgia without Physical Signs This is likely to be of serious significance in young people. Later in life it may be merely a symptom of that type of muscular rheumatism associated with anxiety neurosis, or with chronic fatigue endocrine disturbances, or chronic toxic states [p. 4]

In the young remember three important conditions

1 *The pre-rheumatoid state* the onset of rheumatoid arthritis before the appearance of frank arthritis. The transition from arthralgia to recognizable arthritis is gradual Before the appearance of typical joint signs diagnosis may not be possible. Arthralgia is particularly common as a prelude to the rheumatoid polyarthritis associated with psoriasis. An E.S R., haemoglobin estimation, and X ray films of the hands may be helpful.

2. *The pre-spondylitic phase of ankylosing spondylitis* Arthralgia is one of the several modes of onset of this disease [p. 87] For all adolescent and young adult patients, especially males, complaining of recurrent arthralgia, X ray examination of the sacro-iliac joints should be a routine procedure Candidates for ankylosing spondylitis can be picked out infallibly

3 *Subacute rheumatism.* The importance of its recognition is, of course, that it carries the same liability to carditis as rheumatic fever

Vagrant pains in limbs and joints recur at intervals. They are common in the lower limbs, especially behind the knees [p. 170].

In a child obviously out of health—pale tired and sleeping badly—who has a rapid pulse, a dilated heart, and a cardiac bruit, the diagnosis is obvious. These children have acute rheumatic carditis.

So it is in a child who though apparently in good health, shows clear evidence of old endocarditis—an aortic diastolic bruit, or a presystolic apical bruit, or a systolic apical bruit conducted well out into the axilla.

In a child out of health with no heart signs, it is wise to make a provisional diagnosis of subacute rheumatism, especially if he is subject to recurrent sore throats, if there is a hypochromic anaemia, and if the E.S R. is raised.

Monarticular Onset One knee may be the only joint showing signs of arthritis at the onset of rheumatoid disease. It may remain monarticular for many months, but other joints usually become involved within a year The diagnosis is considered on p 154

I have never seen rheumatoid arthritis remain monarticular for more than a few weeks in any other joint.

Acute Febrile Onset In rheumatoid arthritis an intermittent temperature rising to 99-100° F in the evenings is quite common and some patients are prone to bouts of more severe pyrexia.

Less commonly the disease begins abruptly as a severe febrile illness. A remittent temperature rising to 101 or 102° F lasts for 3-6 weeks and slowly subsides. Joint pain is apt to be severe. When the acute illness subsides, the features of a typical chronic or subacute rheumatoid arthritis persist.

Clinical signs of endocarditis are never found, and there are no electrocardiograph changes

It is usually confused with rheumatic fever Many other disorders, however including some of the other collagen diseases, may resemble it much more closely they are considered in the following section.

ACUTE FEBRILE POLYARTHRITIS

The differential diagnosis is

Acute rheumatoid arthritis

Rheumatic fever

Other causes of polyarthritis migrans allergic joint reactions erythema nodosum purpura rheumatica acute polyarticular gout ulcerative colitis

Collagen diseases systemic lupus erythematosus polyarteritis nodosa diffuse scleroderma dermatomyositis

Hypertrophic osteo-arthropathy

Infective arthritis gonococcal dysenteric meningococcal brucellar

Reiter s disease

RHEUMATIC FEVER

The type case is a child 7 or 8 years old, taken suddenly ill with fever and joint pains though the first attack may come at any age from 3 to 30 years.

A sore throat often precedes the illness usually by some 10 days or more. With the onset of fever a knee or an ankle rapidly becomes swollen, red and exquisitely painful Next day the inflammation may be subsiding, only to start acutely in perhaps a shoulder and a little later a fresh joint is stricken with the same painful swelling. An affected joint returns to normal in at most 2 weeks no permanent stiffness remaining.

The temperature runs a remittent or intermittent course, rarely passing 102° F., for 3 weeks or more. Anaemia quickly develops. Physical signs indicating endocarditis or more rarely pericarditis, may appear at any time, often at the beginning of the second week. Rheumatic nodules, from a pin head to a pea in size, may be discovered on the scalp, or the margin of the scapula or ulna, or on other bony prominences

A febrile illness cannot be rheumatic fever if pain is confined to one joint

One may of course have to decide at once on the possibility of acute osteomyelitis it may be unwise to wait a few days to establish the persistence or migratory character of the joint reaction. In osteomyelitis maximum tenderness is found on percussing the bone of the infected area. But when in doubt, antibiotic therapy is started at once.

A febrile illness cannot be rheumatic fever if pain in a joint remains severe for more than a few days

Many cases of acute rheumatoid arthritis have been so confused.

The flitting polyarthritis of rheumatic fever one joint being attacked as another subsides, the reaction being of short duration—a few hours to a few days—and subsiding with no joint damage persisting, is seen in a number of other disorders. But this is not a source of much diagnostic difficulty Rheumatic fever is more commonly confused with some form of acute febrile polyarthritis not showing a vagrant character

The most important diagnostic characters of rheumatic fever are

1 Maximum incidence in children and adolescents
2. History of previous attacks.
3 Family history of rheumatic fever
4 Tonsillitis some 10 days before the onset.
5 Subsidence of arthritis within about 48 hours under full doses of salicylates

6 Subcutaneous nodules on bony prominences but these occur only in children, and may be absent.

7 Heart signs indicating endocarditis appearing at the beginning of the second week.

STILL'S DISEASE

This is now believed to be rheumatoid arthritis in children most cases begin in the first decade. Sometimes it has an acute febrile onset with severe joint pain, and in these cases there may appear lymphadenopathy splenomegaly a macular rash, subcutaneous nodules on bony prominences and, exceptionally signs of carditis. It is not surprising that an acute febrile case is often mistaken for rheumatic fever But there is one very important difference the polyarthritis of Still's disease never has a vagrant character The joint inflammation is enduring, and does not subside as another joint is attacked

More often the onset is insidious Sometimes it is monarticular the common mistake then is to diagnose tuberculous arthritis. The child loses weight and looks ill, and a typical rheumatoid polyarthritis develops. Anaemia is constant the E.S R. is always raised the Waaler-Rose test is usually negative The prognosis is better than in adults in many cases the disease becomes inactive, leaving a very variable degree of joint damage.

OTHER CAUSES OF POLYARTHRITIS MIGRANS

Allergic Disorders These include serum sickness, giant urticaria, drug oedema, and are associated with joint reactions of this type.

Purpura Rheumatica. This is no longer considered to be related to rheumatic fever It occurs in either sex, aged 10-30 years. Successive and extensive crops of purpuric spots, anywhere on the body but especially on the lower limbs, are accompanied by a migratory severe polyarticular arthralgia, sore throat, and fever

Erythema Nodosum. This affects females more often than males, aged 10-30 years. It is characterized by successive crops of painful, red, tender cutaneous and subcutaneous nodules, usually oval and perhaps several centimetres in length. They come most often on the legs, less commonly on arms and hands. The nodules do not ulcerate they involute in 10-14 days Successive crops continue for 2-6 weeks

Vagrant arthralgia, or polyarthritis resembling that of rheumatic fever is common at the onset, and may precede the erythema nodosum by a few weeks. There is a continued pyrexia, 100-102° F., with sweating, malaise and sore throat.

The E.S R is found increased, often considerably in all cases it

returns to normal as the erythema nodosum subsides The joint reaction subsides completely

The old hypothesis that erythema nodosum is a manifestation of either tuberculosis, or rheumatic fever is no longer accepted It is often found associated with an upper respiratory infection by β-haemolytic streptococci but there is no very good evidence that this is a causal association. The current opinion is that it is a hypersensitive reaction to one of a number of infections, drugs or other allergens

One clearly established association is with sarcoidosis.

The main criteria for the diagnosis of sarcoidosis are

1 Biopsy of skin lesions, or affected lymph gland. Typically there is a general lymphadenopathy Skin lesions—firm, brownish or bluish nodules on face, arm or back, or on the hands at the interphalangeal joints, or a diffuse infiltration on the face—are present in about 50 per cent.

2. The Kveim test an intracutaneous injection of an emulsion of a sarcoid skin nodule is followed by a specific cellular reaction resembling sarcoid tissue.

It has been shown recently[1] that erythema nodosum is frequently a manifestation of sarcoidosis In 24 of 27 cases of erythema nodosum James found bilateral enlargement of hilar glands. In all evidence of sarcoidosis was obtained by a positive biopsy or a positive Kveim test, or both. He points out that at the onset of erythema nodosum careful examination of the skin may reveal sarcoid lesions, which may subside within a week biopsy which may establish the diagnosis, should therefore not be delayed

The conclusion from this and other work is that a more extended use of the Kveim test, in addition to biopsy when possible may show that sarcoidosis is the usual cause of erythema nodosum (but see below under ulcerative colitis)

This appears to be a form of sarcoidosis in which the prognosis, with no treatment is good in James s series, hilar adenopathy disappeared after a year in 64 per cent. and after 2 years in 90 per cent.

Ulcerative Colitis. A migratory polyarticular synovitis resembling that of rheumatic fever may accompany ulcerative colitis the frequency of this complication has been recorded in various reports as from 4-22 per cent. In a series recently published,[2] it is shown that remissions and recurrences may occur over many years the arthritis usually remitting with the colitis, and following an exacerbation of bowel symptoms Residual joint damage is uncommon, but may occur when it does, it resembles that of rheumatoid arthritis.

[1] James, D G., *et al* (1956) *Lancet* ii, 218.
[2] Bywaters, E. G L., and Ansell, B M (1958) *Ann. rheum. Dis.*, 17 169

It is comparatively common for erythema nodosum to accompany a recurrence of this polyarthritis migrans.

Acute Polyarticular Gout This is very rare. It is a succession of attacks of acute gout in a number of joints e.g. first in a foot when this is subsiding, or shortly after subsidence a knee is attacked then an elbow and so on. Successive attacks may continue for about 3 months. There may or may not be a moderate pyrexia

Acute polyarticular gout occurs at an older age than rheumatic fever —above 35 The special diagnostic features are given on p. 118

The arthritis of *polyarteritis nodosa* and of *acute dermatomyositis* and of some *acute infective diseases* may also have this vagrant character [see pp 202, 204 206].

COLLAGEN DISEASES

The collagen diseases are so-called because fibrinoid degeneration of connective tissue, in any or all of the systems, is a histopathological feature common to them all. Another link is that they are all to some extent suppressed by cortisone. As well as rheumatic fever and rheumatoid arthritis, the group includes the following diseases.

Systemic lupus erythematosus frequently resembles rheumatoid arthritis so closely as to suggest that rheumatoid arthritis may be a benign form of systemic lupus. Arthritis similar to that of rheumatoid arthritis may occur in polyarteritis nodosa, but it is far less prominent than in systemic lupus Dermatomyositis and diffuse scleroderma can as a rule be still more clearly distinguished from rheumatoid disease.

Systemic lupus erythematosus

Systemic lupus erythematosus 20 years ago was a rarity today it seems not uncommon—but this may mean simply improved diagnosis. One of the most important recent reviews is that of Hill[1] in the Lumleian lectures of 1957 and I am largely indebted to these papers for the following information.

Modes of Onset The case may be initially benign or initially malignant. The benign form is usually an arthritis of the rheumatoid type, or much less commonly chronic discoid lupus. The two combined suggest that the patient is near a malignant phase.

The commonest mode of onset, in either case, is with joint symptoms a vagrant arthralgia, a polyarthritis resembling that of rheumatic fever or an arthritis of the rheumatoid type either chronic or acute.

Many cases are indistinguishable from rheumatoid arthritis until some unusual feature appears that suggests revision of the diagnosis. A patient of mine a young woman, had a severe polyarthritis of the

[1] Hill, L. C. (1957) *Brit. med. J.*, ii, 655, 726.

rheumatoid type with typical rheumatoid subcutaneous nodules and she developed severely painful deep ulcers on her legs She was therefore tested for the L.E. cell phenomenon, and this was positive.

In a case of apparently typical rheumatoid arthritis the following should suggest that the correct diagnosis is systemic lupus erythematosus

1 the development of severe constitutional symptoms
2. a persisting high E.S R when the joints are quiescent
3 chronic ulcers on the legs
4 evidence of involvement of other systems
   (i) leucopenia thrombocytopenic purpura.
   (ii) pleurisy chronic interstitial pneumonitis pericardial effusion
   (iii) enlargement of lymph glands, spleen or liver
   (iv) renal damage.

Of these ominous signs probably 1 and 4 (iv) are the most common. 4 (i) is especially disconcerting if the patient has had gold treatment a mistaken diagnosis of toxic agranulocytosis is very probable

About 80 per cent begin with joint symptoms the rest with involvement of some other system.

Hill has shown that the well known chronic discoid lupus erythematosus, with its characteristic butterfly wing pattern on the cheeks, is capable of systemic spread—though the risk is small

Pleuritic pain is a frequent early symptom from a pleurisy usually dry or with only small effusion

Thrombocytopenic purpura indistinguishable from the idiopathic disease may precede by months or years manifestations of systemic lupus. It seems probable that in such instances the purpura was the first sign of lupus.

Course of the Disease A malignant phase which may mark the onset, or follow benign symptoms, is one of severe febrile constitutional disturbance. In most cases there is a polyarthritis which may be severe and in addition one or more symptoms and signs of involvement of other systems for instance pleurisy pericarditis, increasing anaemia or blood dyscrasia. A rash—an erythema on face, chest hands, forearms, and indeed anywhere on the body—appears at some time during the course of the illness in approximately 50 per cent of cases Renal damage is serious uraemia is a common terminal event. Neurological and psychiatric symptoms are common severe anxiety depression, hallucinations, mental deterioration convulsions, polyneuritis, hemiplegia, have all been recorded Enlargement of lymph glands, spleen and liver are all common.

80-90 per cent of patients are females the maximum age incidence is between 30 and 50 years.

A remission for a year or more is common and some patients have more than one. But the prognosis even with steroid therapy is bad. It would seem that not more than about 50 per cent. are alive 4 years after the diagnosis is made Many die within a year

Laboratory Findings The diagnostic criterion is in many instances the *L.E. cell phenomenon* The L.E. cell is a polymorph distended by a mass of homogeneous hyaline material which it has engulfed. This is so big in relation to the polymorph that a rim only of cytoplasm is left, and the nucleus is compressed against the margin of the cell [FIG 53] L.E. cells are formed after removal of blood from the body The phenomenon occurs also if plasma from a patient with systemic lupus is added to normal blood a substance present in the plasma peculiar to systemic lupus is responsible for the production of these homogeneous masses, but the nature of the change is unknown. A recommended technique is to defibrinate 5-10 ml of whole blood, transfer to a Wintrobe tube, incubate 2½ hours at 37° C., centrifuge and examine the buffy coat. L.E. cell formation is probably specific but negative results may be obtained in clinically undoubted cases.

Anaemia, normocytic and normochromic, is common. Most cases, but not all, show a polynuclear leucopenia A reduction in the platelet count is fairly common.

The E.S R. is considerably increased usually about 50 mm in 1 hour Almost invariably abnormalities are present in the plasma proteins reduction in the albumin/globulin ratio a slight rise in α_2 and a big rise in γ-globulin, shown by electrophoresis.

Polyarteritis nodosa

This disease is rare. In a recent survey[1] records of 111 histologically proved cases of polyarteritis in 9 teaching centres in Britain from 1946 to 1953 were found 96 of them were submitted to analysis. Much of the following information comes from this report.

It occurs most commonly in men the age incidence shows a steady *rise to a maximum in the 7th decade.*

The morbid changes affect small and medium sized arteries a multiple focal necrosis followed by reactive cellular infiltration. Arteries in any system may be involved the clinical picture is therefore apt to be heterogeneous It is this odd bizarre picture that may suggest the diagnosis it may be hard to think of anything else that could explain the peculiarly diverse clinical findings.

[1] Ross, C. A. (1957) *Brit med J.*, ii, 1148.

As a rule the diagnosis in life is suspected not proved It can be proved only by histological examination of affected tissue this is sometimes possible by skin or muscle biopsies Muscle pain and tenderness is a common initial symptom In Rose s series polyarteritis was demonstrated by muscle biopsy in 40 per cent of cases The skin lesions that may appear are purple mottling in the distal parts of arms and legs tender punched-out ulcers subcutaneous nodules or superficial papules.

A constitutional illness is prominent at some stage in all patients Characteristically this is a painful pyrexia of unknown origin and in a minority the picture is one of acute febrile polyarthritis. The E.S R is high. There is a leucocytosis of 20,000 or more but no discoverable infective agent.

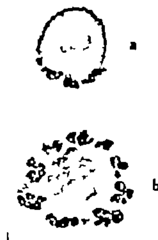


FIG 53

L.E. cells—diagrammatic. (*a*) amorphous mass ingested by a neutrophil (*b*) phagocytosis of an amorphous mass by surrounding neutrophils.

The pain is often abdominal from ulceration of the alimentary tract but it may also be from arthritis, polyneuritis, or polymyositis. Rose found joint symptoms in 27 per cent. 7 patients had a polyarthritis indistinguishable from rheumatoid arthritis 2 had typical hypertrophic osteo-arthropathy

The arthritis of polyarteritis nodosa as a rule is either a polyarthritis migrans like that of rheumatic fever or it much more closely resembles rheumatoid arthritis. Like both, it is suppressed by cortisone Incompatible with both are the many symptoms of systemic polyarteritis.

In Rose s series 32 patients had respiratory illness which preceded the onset of systemic polyarteritis, usually by less than a year but

occasionally by more. They suffered from asthma, or pneumonia, or chronic bronchitis almost certainly manifestations of the specific pulmonary lesions of polyarteritis. At autopsy severe morbid changes resembling those of tuberculosis—fibrosis, nodular and caseous lesions —are found but tubercle bacilli are never demonstrated, while polyarteritis may be.

The larger group of this series remained free from pulmonary involvement.

Renal involvement, shown by proteinuria, microscopic haematuria, granular and hyaline casts, is common and renal failure is a common cause of death. Hypertension develops as a sequel of renal polyarteritis and death may come from heart failure In the group with pulmonary lesions death is more often from the lesion in the lungs.

Diffuse scleroderma

Some women with sclerodactyly quickly develop diffuse scleroderma the connective tissue disease becomes generalized. Men are also affected, and some reports say more frequently

An initial oedema of hands, forearms and face gives place to sclerosis of varying degree sometimes the lower limbs and trunk are also affected It may be only a loss of the normal wrinkling of the face that is seen the textbook picture of a smooth-skinned expressionless face, with a thin tight lipped mouth not opening as it should, and eyes not shutting as tightly as they should, is an advanced stage Not only does sclerosis spread in the skin there is a progressive systemic sclerosis that may affect all connective tissue structures. Symptoms may therefore come from any of the systems.

The diagnosis is based on the spread of a localized scleroderma of hands and feet, and the appearance of systemic symptoms. The change is gradual and is often associated with a low grade fever malaise, loss of weight, arthralgia and myalgia. The painful joints, however can be distinguished from those of rheumatoid arthritis.

The morbid change may affect skeletal muscle, causing pain and stiffness. A myocardial fibrosis may be responsible for myocardial failure Late in the disease X ray films may show a diffuse pulmonary fibrosis In the alimentary tract there may be a patchy fibrosis of mucosal and muscular layers from the oesophagus to the sigmoid and, it may be, oesophageal, gastric or duodenal ulcers. Involvement of the oesophagus and alimentary tract gives an important radiological diagnostic sign loss of the normal oesophageal peristalsis shown by a barium swallow and extreme sluggishness of gastro-intestinal movements. Evidence of renal involvement may also be found. Diffuse

calcinosis is at times observed calcium deposits on fingers, elbows patella, shin, and other parts subject to pressure

Diffuse scleroderma is usually fatal but its course may be very slow Death may come from cachexia, renal or myocardial failure or pulmonary complications.

Laboratory Findings These except for skin and muscle biopsies are not diagnostic. There is a mild anaemia and a moderate leucocytosis There is the reversal of the albumin/globulin ratio common to the collagen diseases

Acute dermatomyositis

This collagen disease is chiefly manifest by a diffuse myositis The early symptoms are fever and malaise, myalgia, and a vagrant arthralgia. It resembles rheumatic fever but there is no response to salicylates, and the diagnosis is suggested by features incompatible with this disease.

It affects females more often than males, and not uncommonly occurs in children.

Usually there is muscular weakness and tenderness and often this affects particularly the shoulder girdle muscles, with resulting difficulty in raising the arms above the head. There may be facial oedema early in the disease and it may further resemble scleroderma in localized fibrotic skin lesions. On the face these may be a butterfly erythema resembling that of lupus erythematosus.

Biopsy of skin and muscle lesions is of considerable diagnostic value if evidence of myositis and of the skin changes of scleroderma are obtained Other laboratory findings—anaemia, slight leucocytosis, increased E.S.R., reversal of the albumin/globulin ratio albumin and casts in the urine—are of little help

The disease is usually fatal within a few years.

HYPERTROPHIC OSTEO ARTHROPATHY

This is a syndrome of 1 clubbing of the fingers and toes 2 chronic periostitis at the distal ends of the long bones 3 arthritis.

The arthritis can easily be taken to be rheumatoid arthritis. Joint pain and tenderness, swelling and restricted movement are at times very severe, and the overlying skin is warm and tender with a brawny oedema. There may be a low grade pyrexia and the E.S R. is increased

It is important to recognize it, for it can be the first manifestation of bronchial carcinoma. It may progress to a crippling degree before lung disease is suspected. It is wise to get a chest radiograph in all cases thought to be rheumatoid arthritis.

This triad is almost always secondary to some chronic disease elsewhere, usually in the lungs. Notably it complicates suppurative conditions, such as bronchiectasis, lung abscess, and empyema. In recent years there have been many reports of its association with bronchial carcinoma, which now seems to be the commonest underlying disease.

There is said to be a difference between cases secondary to chronic suppuration and to lung tumour in the former symptomless clubbing comes insidiously and the arthritis is mild in the latter all symptoms are much more severe and rapid in their development

The syndrome occurs commonly and often severely in cyanotic congenital heart disease and it has been reported in such disparate diseases as Hodgkin s disease affecting mediastinal glands, aortic aneurysm, liver abscess, ulcerative colitis, idiopathic steatorrhoea, chronic intestinal infections intestinal new growth, and polyarteritis nodosa.

The joints chiefly affected are ankles, knees, metacarpo-phalangeal joints, wrists, elbows, According to recent reports, the condition is often treated as one of rheumatoid arthritis for as long as 12 months before a neoplasm is revealed by X ray examination of the chest. The course varies with the primary condition there may be exacerbations and remissions.

When a severe polyarthritis dominates the clinical picture diagnosis *may be very difficult. Pointing to its true nature are clubbing of fingers* and toes, and the X ray appearances of periosteal new bone sheathing the shafts of the long bones in the region of the distal epiphyses. The film may also show porosis of the subarticular bone.

It is a strange disease, the pathogenesis of which is quite obscure. Complete relief of the arthritis very quickly follows operative removal of a lung tumour Patients long stricken with severe pain may within a few days have normally functioning joints again. X ray abnormalities clear up also, though more slowly Not only this symptoms are promptly and completely relieved by severing the vagus on the affected side.

ACUTE INFECTIVE ARTHRITIS

Acute gonococcal arthritis

This disease, very uncommon since the advent of antibiotics, begins 2 or 3 weeks after the onset of acute gonorrhoea. It may be monarticular but two or three joints together may be affected the joints becoming severely painful, hot, red and swollen. The swelling is due to a considerable peri-articular oedema, and a blood-stained turbid synovial effusion. It is an acute pyogenic inflammation the gonococcus can be demonstrated in the joint fluid.

Acute meningococcal arthritis

This may occur during the course of cerebrospinal fever, soon after meningeal symptoms have subsided It is a pyogenic joint infection by the meningococcus tending to leave considerable joint destruction and ankylosis.

Brucellosis (MALTA FEVER UNDULANT FEVER)

Endemic in Mediterranean countries, but rare in Britain There is an insidious onset of pyrexia, headache, and profuse sweats The temperature rises to 103 or 104° F., and the course is a continued fever with a wave-like temperature chart—pyrexia for several weeks, subsidence a second wave of pyrexia briefer and less severe, and so on for some 3 months.

Severe arthralgia is an almost constant complaint in some cases arthritis, often resembling the rheumatoid type, is the prominent clinical feature. The joint reaction, however is often migratory In any continued fever with migratory joint reactions or frank arthritis, diagnostic agglutination tests with *Brucella abortus* and *Brucella melitensis* should be carried out the result of one test is not conclusive, but a rising titre is diagnostic.

REITER'S DISEASE

A triad of non gonococcal urethritis, polyarthritis, and conjunctivitis following bacillary dysentery was described by Reiter in 1916 About the same time, cases of venereal origin were observed

In the last few years it has become increasingly clear that the syndrome may follow gonorrhoea indeed that the usual type of arthritis that follows gonorrhoea is indistinguishable from that of instances of Reiter's disease following bacillary dysentery

We should now be reluctant to diagnose gonococcal arthritis unless

1 the gonococcus is demonstrated in the synovial effusion
2 the type of joint reaction is frankly pyogenic
3 it promptly responds to penicillin.

It seems probable that the venereal form of Reiter's disease is due to an infective agent, possibly a virus, associated with the gonococcus in a mixed infection. True gonococcal arthritis has been seen only very rarely since the advent of antibiotics.

Clinical Features The diagnosis rests on the association of a polyarthritis and a non-gonococcal urethritis, or evidence of prostatitis it derives support from the occurrence of conjunctivitis, or iritis, or a striking skin disorder called keratosis blennorrhagica. Recent figures

for the incidence of these are conjunctivitis 40 per cent., uveitis 10 per cent., keratosis blennorrhagica 10 per cent.[1]

The following information is derived mainly from the accounts given by Harkness,[2] Ford[3] and Csonka.[4]

Women are rarely affected. Csonka's study is based on 185 patients, 182 male and 3 female with Reiter's disease who attended a venereal disease clinic (there is a much higher incidence in women in the post dysenteric cases). The maximum age incidence is 20-40.

Many attacks are closely associated with gonorrhoea. It is often found that gonococci disappear from the purulent urethritis after penicillin treatment but that a residual non-specific urethritis is left, followed by arthritis which is unaffected by penicillin. In some instances infection may be revealed only by examining secretion obtained by prostatic massage. Mason records that, taking 10 pus cells per high power field as the criterion, chronic prostatitis was found in 95 per cent. of Reiter's disease and in 33 per cent of rheumatoid arthritis but the incidence of chronic prostatitis in normal males has been reported as 20-25 per cent.

The gonococcal complement fixation test is positive in about 80 per cent. of patients after gonorrhoea. If therefore a patient showing polyarthritis of this type has chronic prostatitis, a positive gonococcal complement fixation test would strongly support the conclusion that this was venereal in origin.

The joint disease is a polyarthritis of acute or subacute onset, with a tendency to several relapses at very variable intervals. Attacks are of 1 12 months duration, usually about 3 months, and subside spontaneously. They may recur over many years. The main impact is usually on the joints of the lower limb. Small joints of the hands may be involved, as also other joints of the upper limb so may the sterno-clavicular acromio-clavicular and manubrio-sternal joints. Very common and suggestive features are 1 hydrarthrosis of one or both knees 2 acute arthritis of the ankle or tarsal joints 3 tenderness under the heel.

There may be a residual severe pes valgus. A painful calcanean spur from chronic periostitis is peculiarly common. Localized periostitis is seen frequently in X ray films, affecting especially the surface of the calcaneus. There may be permanent changes in the metatarso-phalangeal joints, with local tenderness and pain when walking.

A surprising sequela is the development of ankylosing spondylitis. Mason records that unequivocal bilateral sacro-iliitis was found in

[1] Mason, R. M. *et al.* (1958) *Brit. med. J.*, I, 748
[2] Harkness, A. H. (1949) *Brit. J. vener. Dis.*, 25, 185
(1950) *Non-gonococcal Urethritis*, Edinburgh, Livingstone.
[3] Ford, D. K. (1953) *Ann. rheum. Dis.*, 12, 177
[4] Csonka, G. W. (1958) *Brit. med. J.*, I, 1088.

32 per cent of Reiter's disease and Ford that after several recurrences, typical ankylosing spondylitis may appear

Apart from these two sequelae residual joint damage is uncommon Very rarely a joint may ankylose. The full syndrome is not always seen. Conjunctivitis or iritis may only appear after several recurrences of arthritis and urethritis Iritis is usually late in appearing.

To summarize, the features of Reiter's disease that are of especial diagnostic value are

1 It is an acute or subacute polyarthritis (but occasionally monarticular), (*a*) following gonorrhoea (*b*) following non-gonococcal urethritis (*c*) following dysentery

2. It is associated with urethritis but this may be only slight, and noticeable only first thing in the morning. Results of examination of prostatic fluid may be equivocal much more important is examination of urethral discharge on rising, before micturition.

3 Conjunctivitis, or iritis, very variable in severity occurs in some 50 per cent. of cases.

4 Spontaneous recovery in 3 months or less, with no joint damage, is the rule but the course of the arthritis may be much more prolonged.

5 Joints particularly affected are knees, ankles and tarsal joints.

6 Severely tender points from acute periostitis, especially over malleoli and heel, are common.

Reiter's disease differs from gonococcal arthritis in that the joint exudate is sterile, and the gonococcus cannot be demonstrated in it. Biopsies of synovial membrane have shown an acute inflammation, but it is not a frankly pyogenic joint reaction as is gonococcal arthritis It differs also in that its course is not affected by penicillin.

Reiter's disease differs from rheumatoid arthritis 1 in following a venereal urethritis (gonococcal or not) or diarrhoea (2) in the association with an abacterial urethritis, and frequently with conjunctivitis or iritis 3 in the very common tenderness of the heel from plantar fasciitis and periostitis of the calcaneus 4 in its course.

A prolonged polyarthritis of Reiter's disease may closely mimic rheumatoid arthritis, and diagnosis may be difficult. Neither X ray films nor laboratory tests will help Radiographs in Reiter's disease show no changes other than swelling of periarticular tissues, in the usual acute form If the disease becomes chronic, changes resembling those of rheumatoid arthritis may be found The E.S R. is increased. The Waaler–Rose test is almost always negative.

SPASTICITY

Spasticity means a sustained increase of tone in the limb muscles.

When due to an *upper motor neurone lesion* it is of the peculiar clasp-knife variety the beginning and end of a passive movement are free, but increased resistance is clearly felt in the middle range. It is associated with paresis without muscular atrophy varying in severity in different muscle groups, and the spasticity affects some muscle groups more severely than others this results in characteristic contractures. The physical signs are increased tendon jerks, and if affecting the lower limb loss of the abdominal reflexes, ankle clonus and an extensor plantar response.

The rigidity of *Parkinsonism* unlike that due to a pyramidal lesion, is uniform throughout the whole range of a movement. Passively moving an affected limb feels like bending a soft lead pipe hence the description lead pipe rigidity Cog wheel rigidity is sufficiently descriptive of the modification imposed by Parkinsonian tremor The reflexes are normal

In some cases *hysterical paralysis* is associated with marked spasticity Recognition may be difficult, but the majority will show features never found in an upper motor neurone paralysis. For instance, peculiar contractures develop the plantar reflex is never extensor the patient may be tricked into moving the paralysed limb

If the spasticity is due to an upper motor neurone lesion, the next diagnostic step is to discover its level in the pyramidal system, and if possible its nature

HEMIPLEGIA

A spastic hemiparesis is easy to recognize, unless very slight in degree. The arm is affected more severely than the leg. The limbs assume *characteristic attitudes, which gradually become fixed by contracture* the arm is adducted and flexed at the elbow the forearm pronated, the hand and fingers flexed the leg is extended at the knee, the foot plantar flexed.

The responsible lesion is necessarily unilateral and if above the decussation of the pyramids at the lower end of the medulla, it is on the side opposite to that of the hemiplegia.

The Level of the Lesion. Whether the lesion is above or below the pons is recognized by whether or not the facial muscles are affected.

Facial muscles affected The lesion is usually above the facial nucleus in the pons, and there is weakness of voluntary movements of the face, especially the lower part, on the paralysed side.

A cortical lesion would have to be very extensive to take in leg, arm and face. The lesion is usually in the internal capsule, in the posterior limb of which the whole of the motor fibres for the opposite side of the body are concentrated There may be loss of sensation on the paralysed side if the lesion extends far back in the posterior limb There is often dysarthria from a hemiparesis of the tongue but disorders of deglutition do not occur for the muscles involved receive a bilateral innervation.

If there is a facial paralysis of the lower neurone type on the side opposite that of the paralysed limbs ('crossed hemiplegia') the lesion is in the pons, directly involving the facial nucleus.

Facial muscles not affected. As we proceed from the pons to the cord the pyramidal tracts approximate, and a lesion with a strictly unilateral effect becomes increasingly uncommon. A lesion may reveal its presence in the medulla by causing loss of pain and temperature sense on the paralysed side and by directly affecting cranial nerve nuclei on the side of the lesion.

The Nature of the Lesion. The time for detecting the nature of the lesion is at the onset, or during the initial stage. We are concerned here only with the residual spastic paresis, and will therefore give only a brief outline of the differential diagnosis.

TRANSIENT HEMIPLEGIA. There are episodes of hemiparesis with rapid and complete recovery Elderly arteriosclerotic patients are liable to them, from a transient insufficiency of cerebral circulation. In essential hypertension attacks of severe headache, vomiting, drowsiness, and hemiparesis are liable to occur all signs clearing completely with recovery Lead encephalopathy may closely resemble this. In dementia paralytica epileptiform attacks followed by transient hemiplegia may occur

HEMIPLEGIA IN ADULTS. The commoner causes are

Cerebral vascular events

cerebral haemorrhage cerebral thrombosis	from rupture or occlusion of the lenticulo-striate branch of the middle cerebral artery

syphilitic endarteritis of the middle cerebral artery
embolism of the middle cerebral artery

Cerebral tumour

Cerebral abscess

INFANTILE HEMIPLEGIA. This is a hemiplegia of acute onset in young children. There is a residual spastic hemiparesis, stunting further growth of the limbs. Its origin is uncertain.

SPASTIC PARAPLEGIA

Paraplegia means a paresis of both legs spastic paraplegia indicates a bilateral lesion of the pyramidal tracts

Associated with atrophic paralysis in the upper limbs it indicates a bilateral lesion in the cervical cord sufficiently extensive to interfere with anterior horn cells. A spastic paraplegia with no symptoms or signs in the upper limbs indicates a lesion in the cord below the cervical region—or a very rare bilateral symmetrical lesion of the leg centres in the cerebral cortex A bilateral pyramidal lesion above the cervical enlargement will give a spastic paralysis of all four limbs.

Clearly neurological examination of the upper limbs is essential for some of the data needed for diagnosis

SPASTIC PARAPLEGIA IN ADULTS

The commoner causes of spastic paraplegia in adults, of gradual onset and progression, are

DISSEMINATED SCLEROSIS

CORD COMPRESSION Tumour—meningeal or intra medullary disc protrusion disease of the vertebrae—cervical spondylosis, tuberculosis, new growth pachymeningitis

SYRINGOMYELIA

AMYOTROPHIC LATERAL SCLEROSIS

SUBACUTE COMBINED DEGENERATION (SPASTIC TYPE)

How hard it may be to establish the diagnosis is well brought out in a recent study of spastic paraplegia in middle age.[1] Marshall sets out to determine what becomes of patients diagnosed spastic paraplegia of uncertain aetiology It will be comforting to most of us to know that of the patients he followed up, in roughly one half the diagnosis was still obscure after an average duration of the illness of over 10 years, in spite of very thorough re-investigation by an experienced neurologist. In the rest of the patients re-investigated, a diagnosis could be made. Study of a group of post mortem records showed that the diagnosis could be made in every case after death. In both these groups, *disseminated sclerosis* accounted for about one third of all cases, *cord compression by tumour or disc protrusion* for one third, and the rest were due to *syringomyelia, spinal deformity pachymeningitis* and a number of rarities.

Marshall shows the difficulty of distinguishing disseminated sclerosis from cord compression by a tumour The most helpful points were

[1] Marshall, J (1955) *Lancet*, I, 643

1 cerebrospinal fluid protein above 100 mg. per 100 ml in tumour less than 60 mg. per 100 ml in disseminated sclerosis 2. myelography always abnormal in a tumour

Disseminated sclerosis syringomyelia, and tumour may all present as spastic paraplegia with at first no distinguishing symptoms and signs. In the well established and typical diseases distinction should be easy

Disseminated sclerosis

This is the commonest of the causes of spastic paraplegia. Women are affected rather more often than men, and the disease usually begins in the 3rd decade.

A clue to the diagnosis is often provided by the transient character of the early symptoms. A history of one or more atta ks of *double vision* in a young person is very important associated with pyramidal signs in the legs it makes the diagnosis of disseminated sclerosis almost certain.

Weakness of the lower limb is the symptom for which most patients seek relief. This weakness increases—often with remissions—to a severe spastic paraplegia

Examination of the optic discs shows a pallor of the temporal half in more than one half of all cases.

Paraesthesiae are common There is loss of vibration sense over the malleoli, and there may be loss of postural sense in the lower limbs but there is little or no loss of skin sensation.

Charcot s triad—nystagmus, intention tremor scanning speech—is uncommon, and the diagnosis is usually made in its absence.

Disseminated sclerosis has a much slower course than that of compression of the spinal cord it is further characterized by long remissions, which are absent in compression

Compression of the cord

Clinical features suggesting the probability of a transverse lesion of the cord, such as slow compression, are

1 A history of root pain, preceding the onset of spastic paraplegia. It may be a brachial neuralgia, unilateral or bilateral, or round one side of the trunk, or a girdle pain and it may last for many months.

It should be remembered that the dermatomes on the front of the abdomen range from T 6-7 at the level of the ensiform, through T 10 at the level of the umbilicus, to T 12 just above Poupart s ligament Abdominal pain may therefore be the first sign.

2. Segmental motor signs. These are atrophic paralysis of the muscles

innervated by the segment affected, from involvement of anterior horn or anterior root.

3 Sensory changes in the lower limbs and trunk below the level of the lesion. If there is marked loss of all forms of sensation in the lower limbs, the diagnosis is almost certainly a transverse cord lesion. But the absence of sensory changes does not exclude the diagnosis, for para plegia may appear before sensory loss.

Sphincter disturbances are usually late in appearing.

Changes in the cerebrospinal fluid obtained from the lumbar sac are 1 the protein content is increased to from 100 mg. to 500 mg. or more per 100 ml. (normal 10-40 mg. per 100 ml.) the fluid is yellow from blood pigment, and may clot on standing. 2. a positive Queckenstedt test. During lumbar puncture, with manometer attached to the puncture needle the jugular veins are compressed and released compression normally causes a quick rise in pressure to about 300 mm. of fluid, and there is a rapid fall on release. This does not occur in some 80 per cent. of cases of cord compression the temporarily raised pressure in the cerebrospinal fluid of the cranial cavity cannot be communicated to the fluid in the lumbar sac because of an intervening subarachnoid block.

Grave spinal diseases that may lead to cord compression are those causing vertebral collapse tuberculosis, carcinoma, myelomatosis, osteomyelitis. Severe scoliosis is an occasional cause. Cervical spondylosis is another possible cause among those revealed by X ray examination. If X ray films of the cervical and dorsal spine are negative, the possibilities are tumour intra or extra medullary cervical or dorsal disc protrusion pachymeningitis, softening of the cord from thrombosis of a spinal artery as in syphilitic meningomyelitis.

Tumour and cervical disc protrusion are discussed on p. 77 Cord compression from cervical disc protrusion is less common than from tumour and the duration of the initial root pain is less in disc protrusion. When the protrusion is in the middle line there may be no prodromal root pain. Distinction may however be impossible before operation.

Less is known about dorsal disc protrusion. Only those that compress the cord are operated on, and very few such operations have had to be performed. In one series of 5 500 operations for protruded disc, only 12 were for protrusions in the dorsal region of the spine In all cases severe unilateral root pain was the presenting symptom.

Cervical Spondylosis [p 172] Many people with marked cervical spondylosis shown in X ray films have no symptoms at all. When therefore it is found associated with weakness of the upper and lower limbs, it is not necessarily a causal association.

That it can be so however is now generally accepted Not only may the spinal cord be damaged by cervical disc protrusion it may be damaged also by the much more chronic and very much more common cervical spondylosis (either by direct compression by posterior osteophytes, or by local ischaemia of the cord from compression of the anterior spinal artery)

The symptoms vary according to the nerve tracts in the cord which suffer most. These are usually the pyramidal tracts and the result is a *slowly increasing spastic weakness of one or both lower limbs.* The onset is usually insidious. Occasionally especially after trauma, it may be sudden. There will usually be a history of many months of increasing weakness, and perhaps of cramp and numbness in the lower limbs, of numbness and clumsiness of the hands, and of weakness and wasting in the upper limbs. These symptoms are likely to progress for several years, to a point of serious disability but not complete paralysis then they remain stationary

Syringomyelia [p 16]

Syringomyelia begins in early adult life a progressive muscle wasting, beginning in the hands and spreading up the upper limbs, is associated with loss of pain and temperature sense in the upper limbs and upper thorax. A spastic paraplegia may develop or interference with pyramidal fibres *may be unilateral,* one lower limb only *being affected. It is* a painless condition.

Amyotrophic lateral sclerosis [p. 10]

This is motor neurone disease with the chief impact on pyramidal neurones. It affects typically a middle aged man. It begins with a progressive *atrophy of the small muscles of one hand, which spreads later* to the other hand and then slowly to the arms This is accompanied by a gradually developing weakness and spasticity of the legs. Tendon jerks in both upper and lower limbs are increased there is ankle clonus, but an extensor plantar response is late in appearing.

The condition is painless there is no sensory loss.

Subacute combined degeneration of the cord [p 188]

The spastic type of subacute combined degeneration is the type in which the brunt of the pathological change is borne by the lateral columns posterior columns and peripheral nerves are less affected.

The prominent symptom is an increasing spastic paraplegia. Late in the course of the malady sensory loss and ataxia, indicating degeneration of the posterior columns, appear

The main clue to the diagnosis is the association of nervous signs with pernicious anaemia.

SPASTIC PARAPLEGIA IN CHILDREN

If the child has been able to walk before paraplegia appears, *spinal tuberculosis* [pp 91 105] is the commonest cause

If the child has never been able to walk, the commoner causes are

Congenital spastic paraplegia (*Little s disease*) a condition due to defective development of the pyramidal system. The infant has spastic legs from birth they are rigidly held by muscle spasm in extension and adduction In some instances the arms are also affected (cerebral diplegia) and in these there is associated mental defect.

Congenital defects hydrocephalus, spina bifida with meningo-myelocele

SPASTIC MONOPLEGIA

A pyramidal lesion affecting solely one arm or one leg is uncommon

Spastic paralysis of one arm is likely to be accompanied by weakness of voluntary facial movements on the same side, and by pyramidal signs in the leg of the same side even if the patient complains only of the arm. When strictly limited to one arm the lesion is likely to affect the arm area of the opposite pre-central convolution. Disseminated sclerosis is another possibility

Spastic paralysis of one leg also is likely to be accompanied by pyramidal signs in the arm of the same side, or in the other leg. When symptoms and signs are in fact limited to one leg only the possibilities to be considered are 1 a cortical lesion affecting the leg area of the opposite pre-central convolution 2 disseminated sclerosis 3 syringo-myelia 4 a transverse lesion of the cord below the cervical enlargement, affecting one side only It is in these cases that the uncommon Brown-Séquard syndrome arises on the side of the lesion spastic paralysis of the leg loss of temperature and pain sensation in the opposite leg.

NORMAL VALUES

BLOOD

Cells

RED BLOOD CELLS 4 5-5 5 million per c.mm.

HAEMOGLOBIN 14-16 G per 100 ml (14 G per 100 ml =100 per cent. Sahli)

PACKED CELL VOLUME (P C.V) men, 47 per cent women 42 per cent.

MEAN CORPUSCULAR DIAMETER (M C.D) 7 2 μ

MEAN CORPUSCULAR HAEMOGLOBIN (M C.H.) 29 5 $\gamma\gamma$

MEAN CORPUSCULAR HAEMOGLOBIN CONCENTRATION (M C.H.C.) 34 per cent.

MEAN CORPUSCULAR VOLUME (M.C.V) $86\mu^3$

PLATELETS 200 000-500 000 per c.mm

ERYTHROCYTE SEDIMENTATION RATE (WESTERGREN)

men 0-3 mm in 1 hour
women 0-7 mm. in 1 hour
(Clinically up to 10 mm in 1 hour in men, and up to 15-20 mm. in 1 hour in women should be regarded as unimportant and probably transient.)

WHITE BLOOD CELLS 6 000-8 000 per c.mm.

Neutrophils	60-70	per cent.
Lymphocytes	20-25	per cent.
Monocytes	4- 8	per cent.
Eosinophils	0 5- 3	per cent.
Basophils	0- 2	per cent.

Chemistry (serum or plasma)

CALCIUM 9-11 mg. per 100 ml.

PHOSPHORUS 4-6 mg. per 100 ml. in infancy
2 5-4 5 mg. per 100 ml. in adults.

PHOSPHATASE *acid* 0-3 King–Armstrong units per 100 ml.
alkaline 3-13 King–Armstrong units per 100 ml
alkaline children 10-30 King–Armstrong units per 100 ml.

PROTEIN	total	6 0-8 0 g.	per 100 ml
	albumin	3 5-6 0 g.	per 100 ml
	globulin	1 5-3 0 g.	per 100 ml
	fibrinogen	0 2-0 4 g.	per 100 ml

URIC ACID up to 5 mg. per 100 ml. normal
5-6 mg per 100 ml. equivocal.
over 6 mg. per 100 ml hyperuricaemia.

CHOLESTEROL 150-270 mg. per 100 ml

BASAL METABOLIC RATE

Normal range minus 15 per cent. to plus 15 per cent.

CEREBROSPINAL FLUID

PRESSURE (ADULT) 60-150 mm. water

CELLS 0-5 lymphocytes per c.mm.

PROTEIN 10-40 mg. per 100 ml.

FAECES

TOTAL FAT	10-27	per cent. by weight, of dry faeces.
FREE FATTY ACIDS	2 10	
SOAPS	2 10	
UNSPLIT NEUTRAL FAT	3- 9	

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